# Clinical Orthopaedics


ANTHONY F. DePALMA

*Editor-in-Chief*

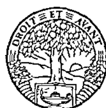

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## ITEMS

# 1

# Michael Hoke

J. Hiram Kite, M.D.*

Michael Hoke was born in Lincolnton, North Carolina, on June 28, 1874. His father was the illustrious Major General Robert F. Hoke of the Confederate Army. His mother was Lydia Van Wyck, of New York. His early years were spent in Lincolnton and Raleigh, North Carolina. After finishing high school in Raleigh, he entered the University of North Carolina. He was athletic, even though he was slender in build and of medium height. He was an outstanding football player and was captain of the team during his senior year. He graduated from the University of North Carolina in 1893 with a Bachelor of Science degree in electrical engineering. The training that he received in mechanics at the university proved to be most valuable to him in later years when he undertook the study of the mechanics of the human foot.

Medicine appealed strongly to the young engineer, consequently, he went to Charlottesville and entered the Medical School of the University of Virginia. He completed the 2-year course and received his degree of Doctor of Medicine in 1895 He interned at the newly opened Johns Hopkins Hospital, in Baltimore. There he came under the influence of Dr. William Osler, chief of Medical Service, whom Dr. Hoke regarded as a very close personal friend. He worked under Dr William Stewart Halsted, who was professor of surgery and the teacher of many famous surgeons. Halsted's teaching and meticulous attention to detail was reflected in Dr. Hoke's surgery in the years to follow. After leaving Baltimore he spent a year at Harvard.

At that time Atlanta had recovered from the War between the States and was one of the most promising cities in the South. Dr. Hoke began practice in Atlanta in 1897, doing general surgery. Later, he specialized in orthopaedic surgery. He was the pioneer orthopaedist in Georgia. He taught orthopaedics at the Atlanta College of Physicians and Surgeons, which later consolidated with the Medical School of Emory University.

On April 20, 1904, after 7 years in practice, Dr. Hoke married Miss Laurie H. Harrison, of Atlanta. The marriage was an unusually happy one: Mrs. Hoke entered fully into her husband's professional life and his many outside interests. They had two daughters.

Dr. Hoke was a member of the Fulton County Medical Association and the Medical Association of Georgia, and he played a prominent part in the founding of the Southern Medical Association. He was a member of the American Orthopedic Association and was elected president in 1925. He presided at the annual meeting held in Atlanta in 1926. This was the only time that the American Orthopedic Association met in Atlanta. When the American Academy of Orthopedic Surgeons was formed later, he was a charter member.

Dr. Hoke's chief relaxations were golf and hunting. He liked the outdoors and pur-

* Atlanta, Ga.



Dr Michael Hoke

chased a large tract of land in South Georgia, where he could go hunting with his dogs. Most of his week ends were spent on the farm. It was not long until he began to experiment with various kinds of grasses for year-round pasture He soon developed a very fine herd of cattle, which won many prizes at the various livestock shows in South Georgia.

Dr. Hoke devoted a great deal of time to working on the charity service of Grady Memorial Hospital

Mrs. W. C. Wardlaw, a prominent citizen of Atlanta, and 6 other ladies sold pencils to raise money for the hospital expenses of a number of crippled children, whom Dr. Hoke treated without pay These were cared for in the Wesley Memorial Hospital, which later became Emory University Hospital. Soon the need of a convalescent home was evident, and 2 cottages were rented in Decatur. As the demands for more beds grew, the ladies were unable to continue to finance the home. At this time, the Scottish Rite Masons of Atlanta, under the leadership of Mr. Forrest Adair, came to their aid, and the Scottish Rite Hospital for Crippled Children was founded in 1915. Operations were performed at Wesley Memorial Hospital for the next 4 years, and the children returned to the home for convalescent care as soon as their condition permitted. In 1919 the present building was built, and it became a complete hospital with an outpatient clinic, operating rooms, x-ray department, physical therapy department and school. Dr. Hoke considered this hospital to be his most important contribution to humanity. He was one of the chief financial contributors to the new hospital and was surgeon-in-chief until his retirement in 1927. It was here that he worked out many of the procedures that were to perpetuate his name.

It was the Scottish Rite Hospital in Decatur that inspired the Shrine of North America to form a chain of Crippled Children's Hospitals over the United States, Canada, Mexico and Hawaii. Seventeen hospitals are now carrying on the work of treating crippled children that was started by Dr. Hoke and Mr. Adair in Decatur. Dr. Hoke was one of the first 5 orthopaedic consultants for the Shriners' Hospitals. One of his stipulations was that they always be kept free of city, state or university involvement.

The highest honor obtainable in Scottish Rite Masonry was awarded Dr. Hoke on October 16, 1923. His patent, hanging in the hospital, states that he received this 33rd Degree "because of the extraordinary services rendered the Scottish Rite in the development of the Hospital for Crippled Children."

Later, Dr. Hoke served for a number of years on the Advisory Board of the Alfred I. DuPont Institute for Crippled Children at Wilmington, Delaware.

In 1931, the University of North Carolina conferred on Dr. Hoke the honorary degree of Doctor of Laws. In the same year, President Roosevelt offered him the post of medical director of the Institute for the Treatment of Infantile Paralysis at Warm Springs, Georgia. It was a difficult decision for Dr. Hoke to make—to give up his private practice in Atlanta and become the full-time medical director of a convalescent hospital that had no operating facilities. Finally, on the President's insistence, he went to Warm Springs. However, he did drive the 75 miles back to Atlanta to do the necessary operations on some of the patients until an operating room was completed at the Foundation. Dr. and Mrs. Hoke lived in the "Little White House" at Warm Springs, vacating it only when the President visited the Foundation. By 1935, Dr. Hoke considered that his work had been completed at the Foundation and returned to his private practice in Atlanta.

From 1914 to 1917, Dr. Hoke worked on a method of stabilizing paralytic feet. At that time various procedures were being reported for this purpose. In his paper describing an operation for stabilizing paralytic feet, he states:

> We have regarded tendon transplantation done alone as unmechanical We have regarded the fixation of tendons to bone to control lateral deformity and lateral mobility as unmechanical, and we have, therefore, done no operations of this nature. We have done no silk insertion operation for this purpose, for we did not believe they would hold against the powerful body weight thrust. We have not done Davis' operation, for it is done blindly and does not take into consideration the architectural details which we think are fundamental. Astragalectomy is certainly an objectionable operation.
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> We believe a stable skeletal foundation is necessary. After paralysis the patient walks on a universal joint over which he has no control. We think it is necessary to do away with the universal joint motion. There is motion in three places. the ankle joint, the subastragaloid joint and the astragaloscaphoid joint. It is necessary to stabilize the subastragaloid and the astragaloscaphoid joints.

Originally, Dr. Hoke did not resect the calcaneocuboid joint, but shortly after 1921 he included this joint, making a triple arthrodesis. His operation differed from the triple arthrodesis of that day by removing the head and the neck of the astragalus and replacing it in different positions according to the different types of deformities in order to restore the normal weight thrust to the bones of the foot. From 1917 to 1921, when he reported his operation, he performed his stabilization operation on 104 cases at the Scottish Rite Hospital.

Dr. Hoke tested a new procedure thoroughly before he reported it. He developed a "clubfoot plastic" operation, in which he cut through the neck of the talus immediately distal to the body and shifted the head medially to restore the normal alignment and weight thrust. Because of the technical difficulty in anchoring the head in the desired position, this operation was not reported.

He was also interested in scoliosis and devised several different types of plaster jackets with a lever and hinge to make pressure on the prominence over the ribs. He also experimented with a derotation table for the correction of scoliosis. This was very much like the table reported about the same time by Galeazzi. Derotation jackets were given up later, and traction jackets were used.

One of his most useful devices was his "well-leg traction apparatus," which was used to treat fractured femurs, to correct flexion contractures of hips, and to pull down congenital dislocation of hips before reduction.

In 1924 he devised and described in the Piedmont Hospital *Bulletin* an operation for recurring dislocation of the shoulder. He drilled a tunnel through the greater tuberosity of the humerus and passed a cord of fascia lata through this and anchored it to the acromion process.

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Dr Hoke was interested in the treatment of cerebral palsy. In 1924, with Dr. Charles

E. Dowman, he published a paper on the treatment of spastic paralysis in which he described the various orthopaedic procedures that could be used in these cases.

Dr. Hoke's mechanical genius is demonstrated best in his interpretation of the mechanics of a foot, as he explained the forces at work in the very relaxed flatfoot. He showed that the anterotibial, the posterotibial and the flexor hallucis longus tendons held up the arch of the foot if the cuneonavicular articulation were stable. Should this joint not be stable, the muscles mentioned had no lever and could not hold up the arch. By fusing this one joint, no stiffness was added to the foot. He performed this operation first in 1923 and reported it in 1931.

In 1937, at the age of 63, he retired from practice in Atlanta for reasons of health and moved with his family to the lovely old town of Beaufort, South Carolina. Their home was known as "Windy Marsh." He died on September 24, 1944.

Dr. Hoke's contribution to the treatment of cripples did not cease with his death; it is perpetuated in the use of the operations that he devised. His inspiration as a teacher continues in the men who had the privilege of working with him; they were instructed thoroughly in the fundamentals of the mechanics of orthopaedics and have contributed much to orthopaedic literature.

# SECTION 1

# RECENT ADVANCES IN ORTHOPAEDIC SURGERY IN INFANCY AND CHILDHOOD

# 2

# The Significance of Growth in Orthopaedic Surgery*

ROBERT B. DUTHIE, CH.M., F.R.C.S.E.†

In the orthopaedic care of children we are particularly concerned with abnormalities and diseases that affect a growing individual. Growth has been defined by Weiss[34] as a developmental increase in the total mass and size of the body. Moreover, growth has to be differentiated from development in which there are maturation and differentiation of tissues and organs necessary for the formation and the completion of the whole individual.

Many orthopaedic abnormalities and conditions are aggravated by or result from growth and its disturbances. It is important to correlate the natural history of the disease process with the growth pattern so that the etiology, the treatment and the prognosis can be defined more accurately. Orthopaedists are in a position to follow and to observe normal growth sequences as well as disease processes in childhood; therefore, they can contribute to this subject which encompasses other disciplines of biology, anthropology, medicine and endocrinology.

The purpose here is to review very briefly the physiologic and the endocrinologic background of skeletal growth and to consider various orthopaedic conditions with the known growth patterns.

## SKELETAL GROWTH

**Method of Studying Skeletal Growth.** Many studies of growth, or development, have been carried out during the past half century, but it is now realized that standardization of methods and expression of data with statistical testing is essential. The significance of results from the more commonly carried out *cross-sectional studies* and from *longitudinal studies* has been clearly described by Tanner[31] and Ellis.[7] Cross-sectional studies are carried out more quickly when measurements of trunk and leg lengths, pelvic and shoulder diameters, etc., are taken from numerous children of the same age group. Although they require a greater number of original cases for accurate analysis, they have provided much information of mean height and other such standards of growth. Longitudinal studies in which similar measurements are obtained from the same child over successive time intervals during growth require less statistical proof and have more significance even in small numbers. From these one can study rate of growth or change; i.e., growth velocity, as well as the various mean standards. But, more importantly, it is this type of study that orthopaedists can carry out.

**Results of Growth Studies.** The oldest

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* This work was carried out while the author was a member of the External Scientific Staff of the Medical Research Council at the Royal National Orthopaedic Hospital, London. He is indebted to Mr. H. J. Seddon, C.M.G , D.M , F.R C S , for help and encouragement in preparing this material.

† Professor of Orthopaedic Surgery, The University of Rochester School of Medicine and Dentistry, Rochester, N. Y.

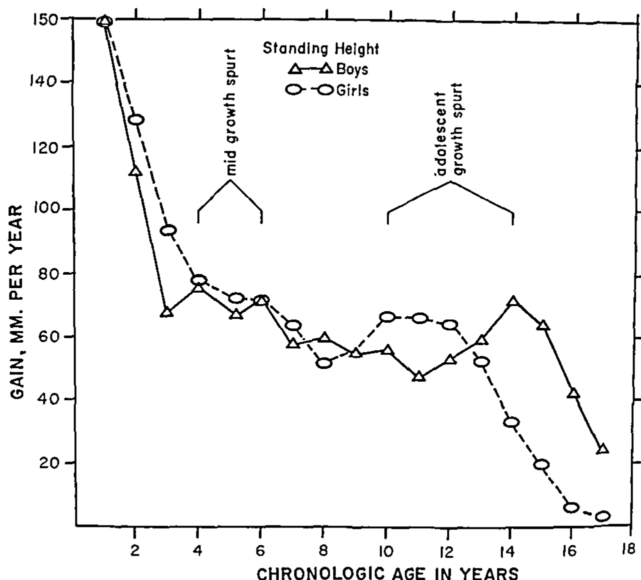



FIG. 1. Showing the standing height mean gain per year in normal boys and girls. (Data from Simmons, K.: Physical Growth and Development, Monogram, Society Research Child Development 9:1)

and most classic longitudinal study was carried out between 1759 and 1777 by de Montbaillard upon his own son.[24] In this the exponential fall in the growth curve is halted twice: firstly, between the ages of 5½ and 7 years, this period being called the *mid-growth spurt* (Tanner[31]), and, secondly, between 13 and 15 years, this being called the *adolescent growth spurt*. This has been confirmed by extensive cross-sectional studies of Simmons[27] and is illustrated in Figure 1. Although much information has been obtained about the adolescent period, relatively little is known about the mid-growth spurt. In orthopaedics many of our problems originate in the mid-growth spurt period or are aggravated by factors that produce it.

SEX DIFFERENCES IN TOTAL STATURE. Although boys may be slightly larger at birth and may grow slightly faster during the first year, between the ages of 1 and 9 years the growth velocities in both sexes are the same (Shuttleworth[26] and Simmons and Todd[28]). The girl at her adolescent growth spurt, which occurs 2 years earlier than in the boy, i e., from 11 to 13 years, grows faster and larger before slowing up to become

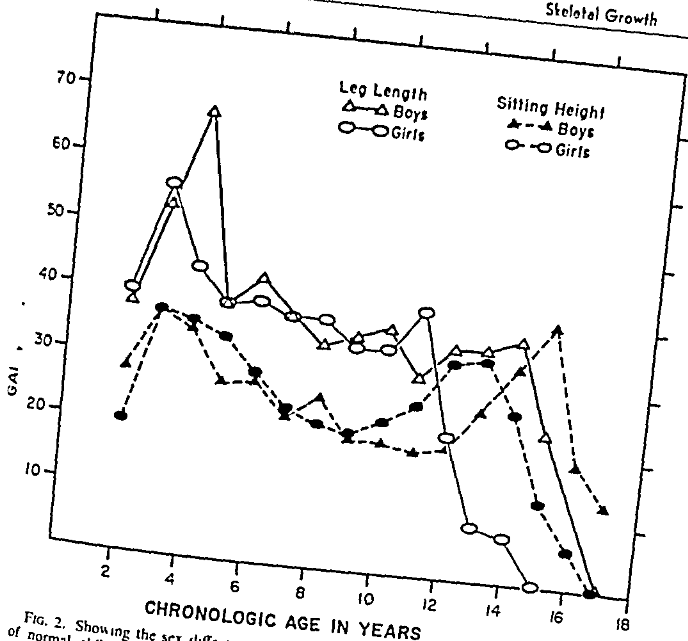



FIG. 2. Showing the sex difference in the leg length and sitting height gains per year of normal children. (Data from Simmons, K.: Physical Growth and Development, Monogram, Society Research Child Development 9:1)

much less in every direction except in pelvic width. The boy then commences his adolescent spurt and grows larger in all directions for a longer period.

DIFFERENTIATION IN GROWTH RATE IN LENGTHS OF TRUNK AND LEG. Total increase in size results from the degree, the duration and the direction of the growth forces. Because of the delay in the appearance of the adolescent growth spurt in boys, there is an extended time for growing and their legs become longer. This time effect is augmented by a differential sex hormone secretion that produces an increased pelvic width in girls and an increased shoulder width in boys.

Accompanying these sex differences in leg lengths and transverse widths is a relative increase in the amount of growth of the trunk or the sitting height to that of the leg (Fig. 2). This is not so marked in girls.

It should be noted that the above data are based upon the chronologic age of the child; i.e., the years since birth. However, marked variation occurs from individual to individual, as well as between somato-type groups.

To reduce this variation, the concept of physiologic or developmental maturity was reintroduced by Crampton.[3] This is based on the time of the appearance of secondary sexual characteristics, of the menarche and of the dental and the skeletal age of the child. The skeletal age is obtained by the aging of a standard roentgenogram, usually of the wrist and the hand, against a wide selection of standard age groupings covering the growing period by Todd[37] or by Greulich and Pyle.[10]

### Factors Affecting Skeletal Growth

#### *During the Prenatal Period*

**Genetic.** These inherited factors may either affect the whole individual primarily by an effect upon the essential metabolic processes of skeletal tissue or secondarily by their direct effect upon the endocrine glands. Crew[4] has shown experimentally that the age of onset of puberty, which regulates the final size, is controlled by genetic factors, but this has not yet been proved in human beings. Genetic studies of families have shown that such conditions as achondroplasia, osteogenesis imperfecta, chondrodystrophy, spina bifida and congenital dislocation of the hip result from a single gene mutation with or without added environmental factors. The genetic influences upon normal somato-type groupings are also well appreciated.

**Maternal Nutritional or Vitamin Deficiencies, Toxemia, Damage or Disease.** Any of these may affect the embryo directly or by changing its essential environment with effect upon its subsequent growth. This is well illustrated by an athyrotic, or hypothyroid, state of the mother, resulting in the birth of a cretin with abnormality of both its osseous development and growth. Although viral infections in the mother during the early weeks of pregnancy have been related to appearance of eye and ear defects in the fetus (Gregg[9]), no skeletal defects have been associated with such infections. Similarly, irradiation to the mother is thought to produce fetal abnormality or malignancy by genetic mutation or by producing direct metabolic changes in the cells. The occurrence of prenatal irradiation, viral infections or threatened abortion has been shown by Stewart, Webb and Hewitt[30] to be considerably higher in children dying of malignancy than among live children. Yet fetal irradiation does not seem to account for the recent increase in malignant disease in childhood.

Duraiswami[6] has demonstrated experimentally in chick embryos the teratogenic effect of insulin, producing numerous congenital defects, particularly of the skeleton. These also can be produced by numerous unrelated agents. However, no human abnormality has ever been related to any similar metabolic disorder, although insulin is regarded as an essential hormone for growth. For many years it has been known that in the East a diet of sweet-pea meal will produce skeletal abnormalities in man[16] and animals,[18] but no such dietary factor has been discovered in the West in humans.

#### *During the Postnatal Period*

**Environmental, sociologic, climatic and facial factors** influence the age of onset of puberty.

**Genetic** factors continue to be operative.

**Nutritional or dietary deficiencies of calcium and phosphorus and vitamins A, C and D** are well-known disorders that produce disturbances of calcification and ossification. Sufficient intake of protein or essential amino acids and iodine is necessary for the functioning of endocrine glands, such as the pituitary and the thyroid, as well as for the formation of tissues during growth It appears that a minimum dietary intake of calories prevents stunting. Diseases of the pancreas, the liver or the kidney can disturb the intake and the output of essential metabolites, thereby producing maldevelopment.

**Infections, Malformations and Vascular Insufficiency Diseases.** Chronic anoxemia from pulmonary or cardiac disease can produce dwarfism. Destruction of motor nerve cells by the poliomyelitis virus will

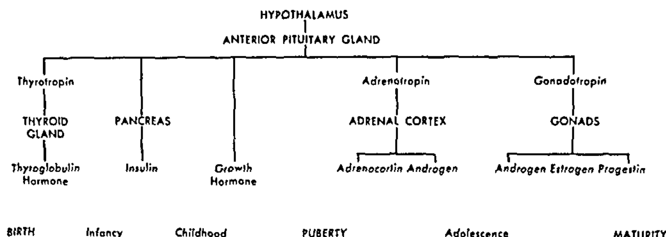



FIG. 3. Illustrating the hormonal factors known to influence the growth of the skeleton.

produce stunting or disturbance of skeletal growth. Ring[21] has shown how the degree of limb shortening is proportional to the extent of the paralysis: there is loss of muscle bulk, with consequent vascular insufficiency, which retards the growth of bone.

**Hormonal.** Until puberty, when there is marked sexual maturation and growth followed by the adolescent growth spurt, there is no real difference in the growth patterns between boys and girls. This "asexual" period of growth is concerned mainly with the anabolism of proteins under the influence of the *growth hormone* of thyroglobulin and, to a lesser extent, of insulin. Their synergistic actions have been described by Hubble[12] and result from secretions by the anterior pituitary gland under the influence of the hypothalamus. During this period the presence of adrenotropin hormone is also recorded, but it is not considered to function until the next phase of growth, when it is responsible for the secretions of androgens and adrenocortin hormones.

At puberty and until maturity there is added to these hormones, which now are gradually diminishing in amount and action, the secretion of gonadotropin from the pituitary. This results in the production of androgens, estrogens and progestin (Fig. 3).

Studies by Wilkins[36] and others have demonstrated the influence of these hormones on the growth and the development of the human skeleton. Many workers have produced hormonally, in vivo as well as in vitro, changes in the time of appearance of the epiphyseal centers and in their subsequent development and fusion. The direct action of hormones on the epiphyseal plate, as well as on growth in general, was reviewed recently.[29] However, there still is much uncertainty as to how, when and where the various hormones act on the skeleton.

Because the growth hormone has no recognizable "target" organ, it is difficult as yet to assess even its indirect actions, although experimentally it has been shown that it directly stimulates tissue synthesis from amino acids.[23] The time of its action is still unknown, although Wilkins[36] has suggested that it does not act until the third or the fourth year, since a hypopituitary dwarf does not exhibit stunting until this time. If this is so, the appearance of the growth hormone may produce the "mid-growth spurt" phase and may produce abnormality of skeletal cells as they first become exposed to its action.

## THE SIGNIFICANCE OF GROWTH IN CERTAIN ORTHOPAEDIC CONDITIONS

### *The Osteochondroses*

These conditions, affecting various parts of the growing skeleton, have been grouped together because of one common feature—they occur in the cartilage of an epiphysis that is undergoing ossification. King[15] and Goff[8] have described in detail from a volu-

minous literature their pathogenesis and possible therapy. Here we are concerned only with the relationship in time of their occurrence to the general growth of the affected child. It should be appreciated that the ages stated in Table 1 are chronologic, rather than physiologic; sex differences in incidence are poor, and, therefore, marked variation must occur. Even so their consideration indicates that

(1) the condition develops shortly after the appearance of ossification in the epiphyseal nucleus—e.g., in Legg-Calvé-Perthes disease at 4 years and in Osgood-Schlatter disease at 11 years;

(2) it occurs during or immediately before either the mid-growth spurt or the adolescent growth spurt—e.g., Perthes' disease occurs during the mid-growth spurt, and Scheuermann's disease is closely followed by the adolescent growth spurt, which may aggravate it;

(3) because of their earlier maturation such conditions might be expected to occur earlier in girls, and this is seen in Scheuermann's disease and Osgood-Schlatter disease. Similarly, the osteochondritic process should stop earlier in girls, and this is commonly found in Perthes' disease.

Although rapid growth is generally regarded as making epiphyses more susceptible to infection and trauma, Goff considered that children who had Perthes' disease exhibited "decelerated growth" However, by relating the clinical onset of the osteochondroses to the growth spurts, one point is clear—they are exposed to intensive growth factors.

Many causes — genetic, infective, traumatic and hormonal—have been suggested, and Figure 3 indicates the various known hormones that influence skeletal growth and, therefore, may affect the development of this condition A deficiency of thyrotrophic hormone has been suggested, and Wilkins[35] describes a dysgenesis affecting all epiphyses as a sign of hypothyroidism. The growth hormone is implicated in that it appears to act at about the time of onset of the mid-growth spurt. The difference in sex incidence is not marked enough to suggest any influence by the gonadotrophic hormones

Most pathologic studies indicate that osteochrondrosis is associated with avascular necrosis of the epiphysis with fibrosis in the metaphyseal region.[11] However, most pathologic studies of the condition are incomplete, having been based upon fragmentary biopsy material. Because of this and its relation to growth pattern, it is suggested that this disease may be produced by a hormonal change, such as the appearance of the growth hormone, causing a generalized abnormal and extensive proliferation of osteogenic cells. A quickly built up nutritional demand by these cells in one epiphysis may be unanswered by the inadequate development or supply of blood vessels because of trauma to that particular epiphysis. This results in differentiation of primary mesenchymal cells into fibroblasts rather than to osteoblasts, and fibrosis occurs in the metaphyseal aspect of the epiphysis. This will then produce further ischemia of the epiphysis and the formation of partial or total avascular necrosis. Hypertrophy and hyperemia of the ligamentum teres has been described, and this may be a compensatory attempt to relieve the avascular lesion.

Although osteochondritic conditions have been described elsewhere in the skeleton, their rarity in comparison with those affecting the spine and the lower limbs indicates that the transmission of body weight and minor stresses are also important contributory causes.

*Osteogenic Sarcomata*

In recent years, critical analyses of the distribution, the histology, the age and the sex incidence have been carried out by Price[19] in carefully collected series of bone sarcomata. The chronologic age and the sex distribution of these tumors will now be considered in relation to the patterns of skeletal growth.

TABLE 1. THE OSTEOCHONDROSES WITH THEIR AGE AT DIAGNOSIS, SEX INCIDENCE AND THE AGE OF APPEARANCE OF THE EPIPHYSES*

| OSTEOCHONDRITIS OF | USUAL AGE AT DIAGNOSIS | RANGE OF AGE INCIDENCE | SEX INCIDENCE | YEAR OF APPEARANCE OF EPIPHYSIS *Male* | *Female* |
|---|---|---|---|---|---|
| Upper femoral epiphysis (Legg-Calvé-Perthes disease) .... | 5-9 | 3½-15 | M > F | 1 | ½ |
| Tuberosity of tibia (Osgood-Schlatter disease) ...... | 12-15 | 10-23 | M > F | 11 | 11 |
| Navicular bone (Köhler's disease) ............ | 3-8 | 2½-10 | M > F | 2-4 | 2-4 |
| Calcanean tubercles (Haglund's disease) ........... | 8½-15 | 8-22 | M > F | 10 | 8 |
| Head of second metatarsal bone (Freiberg's disease) ........... | 10-18 | 10-45 | F > M | 3 | 2 |
| Upper and lower epiphyses of vertebrae (Scheuermann's disease) | 10-21 | — | M > F | 10 | 10 |

* King, E. S. J.: Localized Rarefying Conditions of Bone As Exemplified by Legg-Perthes Disease, Osgood-Schlatter Disease, Kümmell's Disease and Related Conditions, London, Arnold.

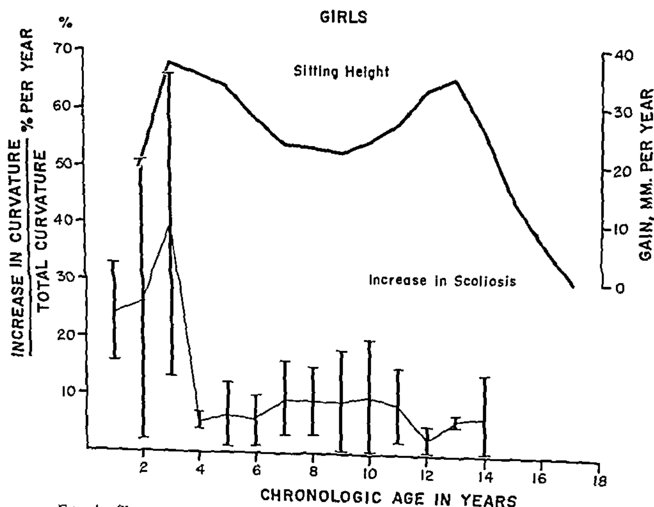



FIG. 4 Showing the mean average increase in the curvature, as a thin line, of infantile idiopathic scoliosis in girls *during* the growing period. The upper graph shows the sitting height gain per year from *Figure 2.*

TABLE 2. AGE AND SEX INCIDENCE OF OSTEOGENIC SARCOMA WITH THEIR NUMBER AND PERCENTAGE IN THE SERIES IN RELATION TO THE POPULATION AT RISK*

| | Years | | |
|---|---|---|---|
| | 0-4 | 5-14 | 15-24 |
| **Males:** | | | |
| Number of osteogenic sarcomata in series.. | — | 25 | 42 |
| % Tumors in series | — | 34.6% | 51% |
| % Population at risk | 13.8% | 29.1% | 30.3% |
| Incidence % tumor in population | — | 1.2 | 1.88 |
| **Females:** | | | |
| Number of osteogenic sarcomata in series.. | — | 24 | 22 |
| % Tumors in series | — | 50% | 45.7% |
| % Population at risk | 12.9% | 28.2% | 30.7% |
| Incidence % tumor in population | — | 1.8 | 1 5 |

* Price, C. H. G.: Primary bone-forming tumors and their relationship to skeletal growth, J. Bone & Joint Surg. 40-B:574-593.

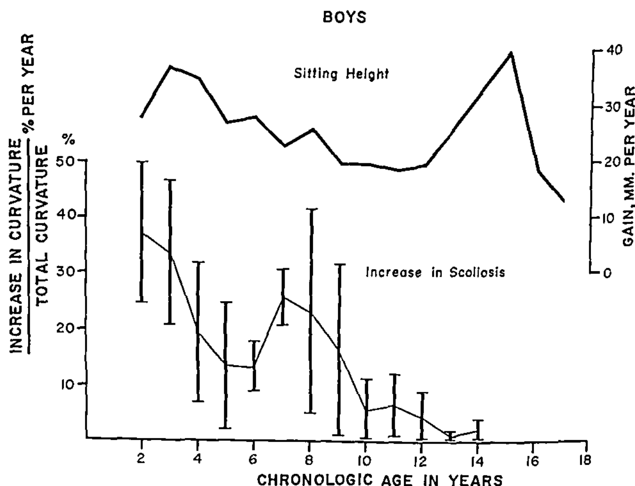



FIG. 5. Showing the mean average increase in the curvature, as a thin line, of infantile idiopathic scoliosis in boys. The upper graph shows the sitting height gain per year from Figure 2

Price[20] has shown from data of the British Empire Cancer Campaign series (1949) and the Bristol Tumor Register series (1949) that the maximum incidence of osteogenic sarcoma in the female occurs between the ages of 5 and 14 years, whereas in the male the maximum incidence is between 15 and 24 years (Table 2). On relating this incidence to the growth pattern, it appears that bone sarcoma develops mainly after the mid-growth spurt in females, but not until after the adolescent growth spurt in males. However, the relative incidence is high in both sexes after the mid-growth spurt, which may imply that the tumor has been present for some time and is aggravated by the factors producing the mid-growth spurt. Up to puberty there is little growth difference between boys and girls; however, there is a difference in the incidence of this tumor. This may result from the earlier onset of the adolescent growth spurt in girls, which results in the earlier appearance of sarcoma. But it is uncertain whether this results from the physical increase in growth or from the change in hormonal secretion, or from both. Unfortunately, the duration of growth of osteogenic sarcoma before diagnosis is not known; therefore, its origin and aggravation by the various growth factors cannot even be surmised. It is of great importance to attempt to relate the appearance of this tumor to the hormonal influences active at that time, especially the growth hormone. This will require an accurate appraisal of the child's physiologic age and status on diagnosis of this lesion and any subsequent change in it during the period of observation.

*Scoliosis*

In order to study the early deterioration of this condition before adolescence, the following clinical review was carried out.

Material. From a very large number of scoliosis patients attending the Royal National Orthopaedic Hospital, London, the records of 10 were selected for analysis. This selection was based on an adequate duration and record of observations of curvatures that had the following properties:

1. Infantile idiopathic type—5 boys and 5 girls.
2. Average extent of primary curve—thoracic 6 to thoracic 12 (range thoracic 4 to lumbar 1).
3. Main curvature to the left.
4. Observed from the age of 2 years (average age of diagnosis 1½ years) until the average age of 12 years (range 10-15 years), when sexual maturity ensued or treatment was begun.

Repeated measurements of the curves on radiographs were carried out using the following method of Ponseti.[17] The upper and the lower limits were identified by choosing intervertebral spaces that were of equal width or wider toward the concavity, with the vertebrae above and below being neutral without rotation. In this longitudinal study some error in measurement may have occurred, but sufficient accuracy was obtained in that all measurements were carried out on standard radiographs over at least 7 years, using the same method. Also, all the radiographs of the same child were available on each measuring.

This particular type of curvature was chosen for study because, although it makes up only about 10 per cent of all cases of scoliosis (James[13]), it is exposed to the total effects of growth throughout its natural history. Because of this it has the worst prognosis, being most progressive and severe of all curvatures (James[14]). In this series, because the same range of vertebrae was involved, this curvature is regarded as being the same entity, whatever its cause and, therefore, should behave in a comparable manner.

Results. The total curvature that developed averaged 97° (57°-144° range), the initial curvature on radiographic diagnosis being 28° (range 14°-47°). The data from this longitudinal study has been expressed in graphic form (Figs. 4 & 5). This shows

the yearly increase in the degree of curvature as a ratio of the total curvature that developed, along the vertical axis, against the chronologic age of the child on the horizontal axis. The total range of all measurements is expressed vertically and the mean average as the central point of this line.

From the present study in both sexes, although of very small numbers and without statistical significance, it is seen that the maximum deterioration occurred up to the age of 4 years. There was a further increase in the rate of deterioration from 6 to 9 in the male child and 7 to 11 years in the female child. There appears to be a more severe deterioration in the male at this age, whereas in the female the deterioration continues for a longer period. The variation in the range at each age is in keeping with the general clinical opinion of marked variation among different children in their age of onset and resulting severity of deterioration. It should be noted that the data charted are based upon the chronologic age of the children, but on correction to their skeletal age this same pattern of deterioration is maintained. The chronologic age was used so that comparison with the mean annual increment figure of sitting height from the Brush Project of Simmons[27] (1944) could be carried out.

Steindler (quoted by Crowe[5]) described the maximum deterioration in scoliosis generally as occurring between the ages of 1 and 2 years, 5 and 10 years, and also at puberty because of the rapid growth of the spine at those times. However, if we consider the yearly increase in the sitting height representing mainly vertebral growth, seen as a broken line in Figures 4 and 5, the initial severe deterioration of the scoliosis curvature occurred at the same period (i e., prior to the mid-growth spurt at 5 to 7 years) There was no definite deterioration associated with the adolescent growth spurt period of 11 to 13 in the female or 13 to 15 in the male adolescent. This suggests that the first deterioration phase accompanies rather than results from the mid-growth spurt. It is more likely that it is aggravated by the factors producing growth as well as the mechanical factors associated with standing. The second deterioration of the curvature may result from the factors producing the mid-growth spurt that it follows closely.

Scott and Morgan[25] also in a small series of cases considered the prognosis of the infantile scoliosis curvature but without any sex difference in its deterioration. They also emphasized the different patterns of deterioration that resulted from differing ages of onset but observed that the rate of deterioration was fairly constant at whatever age it began. It is of interest in the present series that there was little sexual difference in deterioration velocity patterns, and this confirms that up to puberty the skeletal response to hormonal, mechanical and unknown factors is the same. However, the progression in the degree of curvature was not constant, having two peak velocities, when it was related to the total curvature that had developed. At the adolescent growth spurt, the deterioration patterns do differ in the sexes, and this aspect has been dealt with by Risser,[22] Ponseti[17] and Calvo.[2]

It is obvious that for greater definition of this problem it is essential to know the physiologic age and maturity of the child before we can discover the various inherent factors producing this deterioration, their cause or efficient treatment.

## SUMMARY AND CONCLUSIONS

1. The significance of growth to the further understanding of orthopaedic conditions has been emphasized and illustrated by considering the occurrence of osteochondroses, the incidence of osteogenic sarcoma and the increase in the curvature of infantile idiopathic scoliosis.

2 Methods of studying skeletal growth by cross-sectional and longitudinal studies have been defined, the latter method of study being regarded as important in orthopaedic practice.

3. The results of various physiologic studies on growing children are given with a brief description of the various factors that affect the skeleton in the prenatal and the postnatal periods of growth.

4. In a longitudinal study of infantile idiopathic scoliosis, increase in the curvature occurs with two main peak velocities: firstly, soon after diagnosis at the age of 2 to 4 years, and it is related to a growth spurt in the normal sitting height increment; secondly, at the age of 6 to 9 years in boys and 6 to 10 in girls. This peak is not associated with any growth spurt in sitting height but follows closely the mid-growth spurt. The adolescent growth spurt does not appear to have much influence on this particular form of scoliosis, since the maximum deterioration has taken place before this period.

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## Le Signification del Crescentia in le Chirurgia Orthopedic

*Summario in Interlingua*

In patientes pediatric, multe anormalitates e conditiones orthopedic es aggravate o mesmo producite per le crescentia e su disturbationes. Il es importante correlationar le historia natural del processo pathologic con le configuration del crescentia de maniera que le etiologia, le therapia, e le prognose pote esser definite plus accuratemente.

Le configuration crescential pote esser studiate "horizontalmente" per obtener mesurationes ab numerose individuos del mesme gruppo de etate o "verticalmente" per obtener mesurationes ab le mesme individuo a successive periodos de tempore. Le avantages de iste duo methodos as comparate brevemente. Le resultatos de tal studios crescential es describite. In isto, mention es facite del intense crescentia pueritial que occurre inter le etates de 5 e 7 annos e del intense crescentia adolescential que occurre inter le etates de 13 e 15 annos. Le normal differentias in le intensitate del crescentia del statura total e in le intensitate differential del crescentia del trunco e del gambas es comparate, e le importantia del concepto del maturitate physiologic e del etate skeletic es sublineate

Factores que cognoscitemente affice le crescentia skeletic durante le periodo prenatal e alteres que age durante le periodo postnatal es listate In isto, attention special es prestate al factores hormonal.

Tres non-interrelationate conditiones orthopedic es describite con respecto al maturitate disveloppamental e al configurationes crescential que es characteristic de illos. Primo, le osteochondroses—reunite in un sol gruppo proque illos occurre in cartilagine epiphysee durante su ossification—es studiate ab le puncto de vista de lor distribution secundo le etates del patientes al tempore del diagnose e etiam ab le puncto de vista de lor incidentia sexual e del configuration crescential associate con illos. Es constatate que le majoritate de iste conditiones es relationate intimemente con le un o le altere del periodos de intensitate crescential, i e. illo del intensitate crescential del pueritia o illo des intensitate crescential del adolescentia. Es presentate un conception del pathogenese de morbo de Perthes. Secundo, sarcomas osteogene esseva analysate in recente annos con respecto a lor distribution e lor incidentia secundo le etate e le sexo del patientes. Ben que il non existe grande differentias ante le pubertate inter le crescentia de pueros e illo de pueras, le duo sexos non ha le mesme incidentia secundo le etate pro le tumor mentionate. Le incidentia maximal de sarcomas osteogene pro femininas occurre

inter le etates de 5 e 14 annos, durante que in masculos le correspondente incidentia maximal occurre inter le etates de 15 e 24 annos (Price, 1958). Il non es clar si isto resulta ab le facto que le crescentia physic de pueros continua durante un plus longe periodo de tempore o ab alterationes del secretion hormonal o ab ambe iste causas. Finalmente, esseva mesurate le deterioration del curvatura in un gruppo pauco numerose de casos de scoliosis idiopathic infantil. Le studio vertical esseva continuate ab le etate de 2 annos usque al etate—al media—de 12 annos quando le maturitate sexual se declarava o quando le tractamento esseva comenciate. Le mentionate typo de scoliosis esseva seligite proque in illo le curvatura es exponite al effectos del crescentia durante su integre historia natural. Iste studio monstra que le deterioration maximal occurre ante le etate de 4 annos, sequite per un nove intensification del deterioration inter le etates de 6 e 9 annos in pueros e inter le etates de 7 e 11 annos in pueras. Le prime deterioration accompaniava le intensitate crescential del pueritia e non resultava de illo. Il es plus probabile que illo esseva producite per factores de crescentia e etiam per factores mechanic del genere associate con le statura erecte. Le secunde curva de deterioration esseva possibilemente le resultato de factores que produce le intensification pueritial del crescentia le qual illo sequeva directemente.

Il es obvie que un plus precise definition del conditiones orthopedic de patientes pediatric require informationes relative a lor maturitate physiologic e al configuration de lor crescentia. Sin tal informationes nos ha nulle spero de comprender le varie factores que causa o aggrava iste conditiones.

# 3

# Congenital Absence of the Fibula

William D. Arnold, M.D.*

The treatment of a child with congenital absence of the fibula is based upon an understanding of the nature and the usual course of this syndrome. The use of the term *congenital absence of the fibula* is ill suited to the description of the anomaly that bears its name, emphasizing as it does only one aspect of a complex of congenital malformations. These include, in addition to partial absence and complete absence of the fibula, shortening of the extremity, bowing of the tibia and associated anomalies of the foot.

The fibula is the long bone absent most commonly,[10] and the syndrome of congenital absence of the fibula is not infrequent in centers treating orthopaedic disorders. Within recent years there have been 3 reports dealing with approximately 30 cases each of the syndrome. The reader is referred to these sources for detailed descriptions of the deformities and case reports.[4,8,12]

## DESCRIPTION OF ANOMALY

The anomaly presents a characteristic clinical picture in which the following features occur so frequently that one may expect to find all present in a typical example (Fig. 1). These are.

1. Gross shortening of the extremity. In an adult of average size, at least 5 inches of leg shortening may be anticipated. The shortening, while most obvious below the knee, is present to a lesser extent in the femur as well.

2. Anterior bowing of the tibia. The bowing generally exceeds 30° in untreated extremities and usually persists if allowed to remain uncorrected. This contributes to the over-all shortening of the extremity and interferes with walking with or without the use of a prosthesis. The bowing is at the level of the junction of the middle and the distal thirds of the tibia. Occasionally medial bowing is present as well.

3. Equinovalgus deformity of the foot. This is characteristic and persistent. In untreated examples the heel may be posterior and parallel to the axis of the distal tibia.

4. Absence of one or more metatarsal rays.

5. A skin dimple over the apex of the tibial bow in over 50 per cent of the cases.

In addition, congenital anomalies elsewhere are found with increased frequency in this group of patients. Not unusual are associated developmental defects of the upper extremities Bilateral congenital absence of the fibula is present in about 30 per cent of the cases. (When unilateral, the right side is involved more frequently.)

Coventry and Johnson have proposed a classification based upon extent of deformity and associated anomalies.[4] However, it would seem best to consider congenital absence of the fibula in terms of a gradient of deformities ranging from an extremity with

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partial absence of the fibula, 1 to 2 inches of shortening, a straight tibia and a normal foot to a severely crippled limb marked by a partial femoral absence, severe tibial and foot deformities and associated anomalies in other extremities.

The roentgenographic picture of the anomaly is also characteristic. In addition to the absent or hypoplastic fibula, the bowed tibia is clearly visible with a thickened cortex on the concavity of the bow. In contradistinction to the pre-pseudarthrosis state of congenital pseudarthrosis of the tibia, the medullary canal is present. The distal tibial epiphysis is small and irregular; often this is true of the upper tibial epiphysis as well. There may be hypoplasia of the patella and various anomalies of the femur ranging from simple minimal hypoplasia (most frequently) to absence of portions of the bone. Usually there is fusion of the talus and the os calcis and frequently the navicular as well into one bony mass. The cuboid may either be absent or also incorporated into the tarsal bone mass. Nearly always there will be absence of one or more metatarsal rays, and frequently anomalies of the toes are present. Ossification centers are delayed in appearance throughout the extremity, and it cannot be stated with certainty that the fibula or any of the tarsal bones are absent until after the fifth year (Fig 2, *left & center*).



Fig. 1. Congenital absence of the fibula. A severe but typical example of the untreated deformity in a 12-year-old boy. A satisfactory end-bearing stump was obtained by a Peabody osteotomy followed by Syme's amputation.

Anatomic studies of this anomaly are few, but they have provided valuable information in developing a plan of treatment.[7] The important finding, confirmed several times at surgery, is the presence of a definite stout thick fibrous or fibrocartilaginous band of tissue continuous with the lateral border of the intermuscular septum and probably representing the anlage of the absent fibula (Fig. 2, *left & center*). The band extends from the upper tibia to attach to the lateral and posterior margin of the os calcis. Often it adheres closely to the distal tibia. In many patients the band may be palpated as a taut structure lateral to the heel cord. The peroneal muscles and the extensor hallucis longus arise from the band. The peroneus brevis may be absent, but generally there are no gross abnormalities of the nervous or the vascular structures of the limb. In the foot, when rays are absent, the remaining lateralmost ray has been found to correspond with the fifth toe, normal intrinsic musculature of that toe being present.

The etiology of congenital absence of the fibula is not known. There is little to suggest a hereditary factor in most of the recorded cases. Normal embryologic studies point to a developmental accident to the



FIG. 2. (*Left*) Dissection of specimen with congenital absence of the fibula In this leg the tibia was straight. The relatively small band is indicated by the arrow. Note the single peroneal tendon (peroneus longus). Although there are only 4 toes, the lateral ray is provided with the normal intrinsic musculature of the fifth toe. (*Right*) Specimen cleared further (lateral and posterior views). Note the relationship of the band to the interosseous membrane and its distal attachments to the os calcis and the tibia The talus, the os calcis and the cuboid are present as a single tarsal bone mass.

embryo at about the fifth or the 6th week of intra-uterine life. Similar anomalies have been produced experimentally by a variety of means. For the results of these studies, the reader is referred to the classic experiments of Bagg[2] and also to those of Warkany[14] and Duraiswami[5] Attention is also directed to the observations of Freund[6] and Middleton[9] with respect to the etiology of this syndrome

## TREATMENT

The aim of treatment of a patient with congenital absence of the fibula is to provide an extremity equal in length to its mate and suitable for plantar weight-bearing. The importance of correcting the position of the foot to allow weight-bearing on tough plantar skin cannot be overemphasized. All treatment must be individualized in view of the range of deformities that may exist. However, certain methods have proven to be of value and may be adapted to the majority of cases. For the purposes of this discussion it is easiest to consider the therapy of each important component of the anomaly separately.

### TIBIAL BOWING

The tibial bowing must be corrected if it is so severe that it prevents plantar weight-bearing. In addition, it is a factor in producing leg-length discrepancy. Thompson *et al.*[12] first advocated excision of the tight band that acts as a bowstring increasing and preventing spontaneous correction of the

Fig. 3. Unilateral congenital absence of the fibula before and 14 years after removal of the tight band. Complete correction of tibial bowing and equinovalgus deformity. Leg-length discrepancy remains. (Thompson, Straub & Arnold: J. Bone & Joint Surg 39-A:1229-1237)









Fig 4 Bilateral congenital absence of the fibula 6 months and 15 years after removal of bilateral tight bands Correction of tibial bowing and equinus deformity. Fixed valgus due to tarsal anomalies does not interfere with full activities. Leg lengths are equal



FIG. 5 Operative photograph of a rather large fibrocartilaginous band in a 5-year-old boy (Lateral view, heel in lower right of photograph)

deformity. This will permit correction of the tibial bowing within a few years and, as well, release the equinus deformity of the foot (Figs. 3 & 4). As with any other deforming structure, early release is important, and the operation should be performed within the first year of life. It is unlikely to be of much benefit if delayed beyond the age of 5 years Although the band is not invariably associated with tibial bowing, it is the author's experience that a tight band has been found when searched for in every leg with a typical deformity.

The technic of the operation is simple. An incision is made immediately lateral to the heel cord. It is desirable that the skin incision be curved or S shaped to prevent any recurrence of equinus due to skin contracture. Frequently the band will be apparent as a thick structure approximating the heel cord in size (Fig. 5). If not, a little dissection will reveal it beneath the peroneal musculature forming the lateral boundary of the intermuscular septum. The band should be excised as high as possible down to its insertion into the posterolateral portion of the fused talocalcaneal bone mass. If equinus deformity of the foot persists after removal of the tight band, the tendoachillis should be lengthened and, if necessary, a posterior capsulotomy of the ankle performed. Postoperatively the leg is immobilized until soft-tissue healing occurs (2-3 weeks), following which plantar weight-bearing is encouraged. The evidence now is sufficient to state that removal of the tight congenital band is an essential feature of the management of most early cases of congenital absence of the fibula. The only apparent exceptions would seem to be those patients whose legs are straight with feet in a satisfactory position for weight-bearing Severe degrees of leg shortening or partial femoral absence are not contraindications to this procedure, since further therapy for this condition will require a straight tibia and a well-aligned heel.

Correction of the tibial bowing in older children can best be accomplished by osteotomy of the tibia. Release of the tight posterior calf structures, including the congenital band, will be necessary to permit correct positioning of the foot. Osteotomies of the tibia heal satisfactorily in congenital absence of the fibula in contradistinction to the prepseudarthrosis state. In correction of severe anterior and medial bowing of the tibia, the swivel osteotomy described by Peabody may be employed [11]

### LEG-LENGTH DISCREPANCY

In spite of early removal of the tight band, serious leg-length discrepancy will occur even though the tibial bowing has been corrected. In bilateral instances of congenital absence of the fibula, this will not be a major problem provided that the over-all shorten-

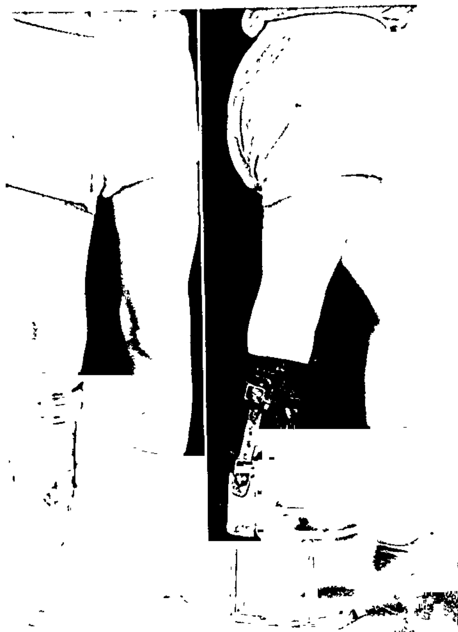


FIG. 6. Patten brace used during childhood. Syme's amputation will be recommended when growth ceases. (Thompson, Straub & Arnold: J. Bone & Joint Surg. 39-A:1229-1237)

ing is not such as to be dwarfing and that the abnormalities of the feet are not so severe as to make weight-bearing impossible. Indeed, in these cases, no treatment other than correction of the tibial bowing and equinus deformity may be necessary, and these individuals may be fortunate enough to lead essentially normal and active lives.

Since 5 inches or more of leg-length discrepancy may be expected in the typical unilateral example of congenital absence of the fibula, a serious problem in therapy is posed. In most cases the discrepancy will be too great to permit correction by epiphyseodesis or femoral shortening or bone-lengthening procedures. Although theoretically a combination of epiphyseodesis and femoral shortening would equalize leg lengths, this cannot be recommended for the average patient. However, in some patients with partial absence of the fibula, the shortening expected would be less, and here the usual procedures to equalize leg lengths would be applicable.

In the typical case, either an elevated shoe or an amputation is necessary. Every effort should be made to start the child walking at an early age. The patten brace illustrated is useful throughout childhood if the ankle is unstable (Fig. 6). In the majority of cases a Syme amputation is preferred. This permits use of plantar heel skin for weight-

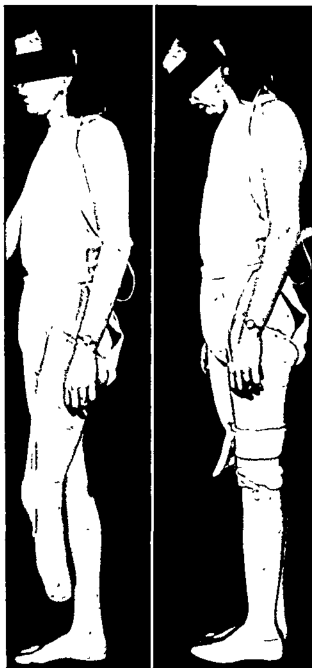


FIG. 7. Syme's amputation performed for congenital absence of the fibula. Note that the end-bearing stump is smaller and falls at a higher level in these patients than in the usual Syme amputation, permitting a better functional and cosmetic prosthesis. (*Thompson, Straub & Arnold:* J. Bone & Joint Surg 39-A:1229-1237)

bearing, and, since the extremity is short, the normally bulbous stump falls at the mid-calf level, permitting a cosmetically acceptable prosthesis even in a girl (Fig. 7). The stump formed is far preferable to a standard below-knee amputation stump. It is necessary that tibial bowing and severe equinus deformities be corrected before a Syme amputation is performed, and it is for these reasons that measures to correct these deformities are urged in infancy even if later amputation is contemplated. Although it has been suggested that the Syme amputation be performed early in childhood,[8] it is probably best to delay amputation for most children until adolescence. The author has seen 2 Syme amputations performed in young children for congenital absence of the fibula that required revisions. However, especially in girls, when psychic factors predominate, consideration should be given to amputating a severely shortened extremity at an earlier age. One may accept the possibility of revising the amputation at a later date in order to provide a cosmetically acceptable prosthesis during childhood. In performing a Syme amputation in young children, the distal tibial epiphysis must be preserved. The problems of a juvenile amputee are many: bony overgrowth, inadequate soft tissue about the stump, and the necessity for repeated prosthetic revisions may be mentioned. The expense of the several prostheses alone may dictate delay in amputation until cessation of growth. For a more complete discussion of this important and complex problem the reader is referred to the publications of Aitken and Frantz[1] and the review by Chittenden.[3]

In the past, other methods were employed to deal with the leg-length discrepancy encountered in these patients. Among them may be mentioned the use of prostheses with the foot held in equinus and various types of ankle fusions with a full equinus foot. Usually these methods are inferior to a Syme amputation. In addition to the clumsy apparatus required, painful weight-bearing has been reported. It is apparent that one of the essentials of treatment — plantar weight-bearing — is not obtainable by the use of these procedures.






FIG. 8. Bilateral congenital absence of the fibula with severe femoral defects. Resection of both tight bands has permitted full plantar weight-bearing on the left and facilitated prosthesis fitting on the right. Note associated defects in the right hand.

### ANKLE INSTABILITY

Ankle instability in these patients is often not as great as might be expected with an absent lateral malleolus. Some patients do not require any support, while most can be supplied with a molded leather steel-reinforced ankle brace fitting inside a stock shoe. The narrow foot and ankle incident to the anomaly allow for easy fitting of the brace inside a shoe. Arthrodesis of the ankle may be indicated in the unusual case, but anomalous fusion of the tarsal bones in a valgus position almost always eliminates the need for arthrodesis of the foot, since the position is satisfactory for weight-bearing provided that the equinus deformity has been corrected. Measures to provide a lateral malleolus by bone grafts have been proposed, but it is believed that the operation seldom is indicated.

### EQUINOVALGUS DEFORMITY

Correction of the equinus deformity has been discussed. Removal of the tight band and, if necessary, tendo-achillis lengthening with posterior capsulotomy of the ankle will usually suffice in young children. Tendon transfers to correct the valgus position of the foot have been carried out on several occasions. In selected patients, transferral of the peroneus longus to the anterior tarsal region may be of value. Generally this is not indicated, since the valgus position is fixed and suitable for weight-bearing if the foot is to be preserved. In addition, one should bear in mind the possibility of congenital absence of one of the peroneal tendons.

Rarely, for extreme valgus preventing suitable plantar weight-bearing, a medial tarsal wedge resection of the usually fused anomalous tarsal bone mass may be required. It should be emphasized that the rigid valgus foot in these patients produced by the congenital tarsal fusion is a surprisingly good weight-bearing structure. One of our patients with bilateral deformities has played football and even won a local high-jumping contest. He has had no treatment except excision of the tight bands to correct the equinus deformities and tibial bowing. Today, 21 years after his operations, he works standing at a laboratory bench, 10, and even 14, hours per day without discomfort (Fig. 4).

Wedging plasters and manipulation by themselves have little place in the treatment of congenital absence of the fibula. Unfortunate conversions of a valgus foot to a varus position have occurred in the presence of a tight band and heel cord. Following removal of the tight band and release of any other tight posterior calf structures there is little tendency to recurrence of the deformity, particularly if plantar weight-bearing is instituted as soon as soft-tissue healing is complete.

### Femoral Defects

Not infrequently femoral defects of considerable degree coexist in these patients and greatly modify treatment (Fig. 8, *left & right*). Turn-up plasty and rotation-plasty have been proposed as means of handling these formidable problems. Van Nes' rotation-plasty could be adapted to certain of these patients when severe femoral defects existed.[13] Since the ankle joint falls at the knee level of the opposite extremity in these patients, Van Nes recommends fusion of the knee and rotation of the leg through an arc of 180° A standard above-knee prosthesis can then be used, the reversed ankle joint controlling the "knee" joint of the prosthesis. Experience with epiphyseal transplants from the opposite normal fibula to replace femoral or other long-bone defects has not been sufficient to recommend this procedure. In some instances, when the bony defects are great, ischial weight-bearing prostheses have been necessary. However, whenever possible, even in the most severely deformed limbs, plantar weight-bearing within the prosthesis should be achieved.

### SUMMARY

It is obvious that the treatment of a patient with congenital absence of the fibula is an individual matter, depending largely on the extent of the various deformities and on the associated anomalies. No brief description can cover all the possibilities. Nevertheless, the following features of treatment require emphasis:

1. Patients with bilateral anomalies have a more favorable prognosis, since leg-length discrepancy is not a problem. Amputation should not be carried out in these patients unless severe deformities of the foot prohibit weight-bearing or the dwarfing is such that increase in height with prostheses is advisable.
2. Removal of the tight band at an early age, permitting spontaneous correction of the tibial bowing, is recommended. This allows plantar weight-bearing in bilateral cases and facilitates performance of a Syme amputation if one is indicated later.
3. The Syme amputation is preferred in most patients with congenital absence of the fibula when amputation is indicated.

In spite of the wide gradient of deformities present in this anomaly, it is remarkable how consistently the same patterns recur. The adult appearance can generally be predicted in infancy when a typical anomaly is present, permitting a rational and consistent program of therapy Of equal importance, the physician is able to furnish sound advice and give the parents an accurate prognosis on these unfortunate children.

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## Le Tractamento de Absentia Congenite del Fibula

*Summario in Interlingua*


Le absentia congenite del fibula es un ndrome que consiste usualmente de acrtamento del gamba, absentia del fibula, rvatura del tibia, deformitate equino-valgal :l pede, fusion del ossos tarsal, e absentia : un o plure radios metatarsal. Le formas :l tractamento varia secundo le grado de veritate del condition, sed le objectivos imari es le equalisation del longor gamba le effectuation de un localisation del ıpportamento de peso in la area del plus ›rte pelle plantar del calce e del pede. In resection del congenite ligamine tense ue representa le anlage del fibula absente esulta usualmente in le correction sponanee del curvatura tibial e in plus relaxa : deformitate equin del pede. Iste inter'ention chirurgic deberea esser effectuate durante le prime anno del vita. Un accurtamento del gamba per plus que 5 pollices es a expectar in le caso typic mesmo post le correction del deformitate equin e del curvatura tibial. Le amputation de Syme es recommendate in iste patientes e deberea esser effectuate post le cessation del crescentia. Amputation non deberea esser effectuate in casos bilateral de absentia congenite del fibula, excepte si sever deformitates del pede prohibi le supporto de peso o si le nanification es si extreme que un augmento del statura per medio de prosthese pare indicate. In despecto de un extense gamma de deformitates occurrente in iste syndrome, certe definite configurationes recurre, e le apparentia del patiente al etate adulte pote usualmente esser predicite in su infantia.

# 4

# Management of the Juvenile Amputee*

Charles H. Frantz, M.D., and George T. Aitken, M.D.†

Children with an amputation are a small but integral part of the crippled children in the population. However, if one adds congenital deformities of the extremities that may be treated as amputations, the group is a sizable one. In the past these patients seldom had the careful evaluation and programming of care accorded the victims of postpoliomyelitis and cerebral palsy.

The extensive work that has been done recently in the areas of modern prosthetic design and fabrication technics under the aegis of the Prosthetics Research Board allows both physicians and prosthetists to manage amputees more ably. It is possible now, with a well-organized program, to fit and rehabilitate practically all types of traumatic and congenital amputees. Of course, the degree of rehabilitation depends on the age of the patient and the extent of the handicap. In most cases, much more can be anticipated today than was the case several years ago.

The increased mechanization of our times has brought our children most of the traumatic episodes that produce amputations: automobiles, railroads, power tools (both in the home and on the farm), plus civil gunshot wounds, account for an increasing number of severe injuries to children that may produce amputations (see following table).

Six-year Survey:
128 Noncongenital Amputees

| 68% Traumatic Origin: | |
|---|---|
| 44% | Vehicular<br>Power tools<br>Railroad accidents |
| 24% | Explosions<br>Civil gunshot wounds |

## THE AMPUTEE TEAM

A well-organized amputee clinic must have available the services of a surgeon, a physical therapist, an occupational therapist and a qualified prosthetist. The services of a medical social worker, a psychologist-psychiatrist and a pediatrician are desirable but not mandatory (Fig. 1).

The rehabilitation of an amputee is the development of a well-adjusted man-machine combination. The man must be properly prepared (surgery and psychology); the machine must be well and comfortably made (prosthetist). Then the two must be joined (fitting) and trial-tested for function. Once comfortably joined, the man must be taught to use the machine to its best advantage (occupational therapy, physical therapy and/or vocational training).

This schema is an oversimplification of an exceedingly complex problem. However, it does demonstrate the role that each mem-

* This work is made possible in part by the cooperation of The Michigan Crippled Children Commission, Carleton Dean, M D , Director.

† Medical Directors of the Area Child Amputee Center, Mary Free Bed Children's Hospital and Orthopedic Center, Grand Rapids, Mich.

ber of the team plays. Each is important and necessary. Because the result is dependent on the best efforts of people in different disciplines, it is necessary that each member of the team be competent, diligent, interested and willing to accept suggestions one from the other for the total benefit of the patient.

Management of the juvenile amputee requires that one recognize clearly some of the areas in which a child is different from an adult. Children are members of family groups. Except within personality limits, their habits and performance are directed by the will of the parents. Children are incapable of desiring an artificial limb on the basis of function. Further, they are not mature enough to discipline themselves to practice use of the limb necessary for the development of good function. Therefore, it is necessary for the amputee team to be aware of these facts in the preliminary evaluation of a young patient. Unless the family is convinced that prosthetic restoration is desirable and advantageous, no significant result will be obtained. Good rehabilitation depends in large measure on the authoritarian demand of the family that the child wear the limb and become proficient in its use.

Children are different from adults in that they are growing organisms. Until maturity they are in a constant state of physical and emotional flux. Their motor skeletal systems are increasing in skill and size. Growth produces problems both as regards repair and replacement of the prosthesis, as well as problems in surgical planning. The mental-emotional flux manifests itself in the areas of performance. A five-year-old amputee cannot reasonably be trained to walk with the grace and the facility of a twelve-year-old. Such variables in training and evaluation are present constantly. They must be recognized by all the team members, and the prescription-writing, the training program and the follow-up plan should be adjusted accordingly.

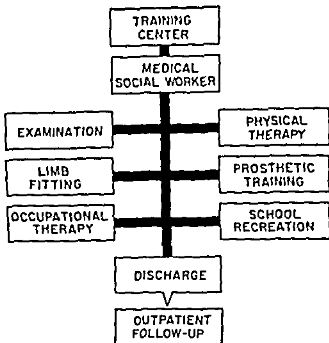



Fig. 1. (Frantz, C. H.: M. Times 87:624-625)

Child amputees are classified in three groups: (1) noncongenital (postsurgical and post-traumatic); (2) true congenital amputations; and (3) appendicular abnormalities.

## AMPUTATION SURGERY

Surgery of amputation in children falls into two major classes:

### 1. Emergency Amputation Surgery

Etiologic factors contributing to this group include acute trauma, necessitating the immediate removal of a limb. Explosions, vehicular accidents, power-tool and railroad accidents lead the list. Cases falling within this definition must be evaluated carefully as individual problems. Trauma producing lesions of a degree requiring amputation usually implies more than local damage. One must exercise caution when confronted with a child whose extremity is severely damaged. If the survival of the extremity is in doubt, every effort must be expended to save it. Delayed amputation is to be preferred to unnecessary amputation.

### 2. Elective Amputation

**Nonfunctional Extremities.** The leading etiologic factors are subacute infection

(gangrene following fracture, etc.), tumors, thermal injuries and nerve injuries. Cases falling within this group may be approached with more deliberation. The nature of the etiologic factors permits more careful evaluation and planning.

The subacute infection complicating a compound fracture, in which gangrene may develop quickly, may be classified in the emergency amputation group. The sequelae of chronic infection in children, definitely less frequent in the past two decades, may present problems from time to time. Severe deformities resulting from long-standing burnt-out osteomyelitis may come to amputation for improvement of function. The repeated saucerization of long bones with invasion of epiphyses and joints is manifested by extensive scarring, loss of muscle, ankylosed joints, chronic draining sinuses and extreme degrees of shortening. The degree of shortening and functional loss will influence the decision to amputate.

Amputation is required in the treatment of many types of malignant bone tumors. The tragedy of a malignant bone tumor in a child can be surpassed only by a mistake in diagnosis. Thorough study and consultation are advisable, utilizing all the facilities of the laboratory and roentgenography to aid in the decision.

Third-degree burns and carbonization may be indications for amputation. In extensive fibrosis of muscle, loss of ankle-joint motion, adherent tendons, loss of plantar skin and epiphyseal damage, function will be better and the future economically will be brighter with a below-knee amputation. The freezing of an extremity that results in gangrene is a primary indication for amputation in cold injuries.

## REVISIONS OF THE STUMP

**Revision of Old Stump.** Overgrowth; neuroma, trophic skin changes

*Revisions of stumps in children are done* primarily for overgrowth, bursae and neuromata. Overgrowth is the major complication of amputations in children. It is a disproportion between the length of the bony stump and the soft tissue. Its mechanism is not clearly understood. It occurs in the fibula, the tibia, the humerus and the radius and the ulna, in that order of frequency (Fig. 2). It is not the result of the *vis a tergo* of the next proximal epiphysis It manifests itself clinically by increasing tenderness over the bony tip of the stump, followed by protrusion through the skin and erosion with formation of granulation tissue.

The treatment for this condition is revision of the stump, resection of the bone at a higher level, or extirpation of the fibula extraperiosteally (Fig. 3). This complication ceases at the time of skeletal maturity

Bursae develop frequently between the bony stump and the end of the soft-tissue stump. This is more frequent in the exceedingly active below-knee male juvenile. Whether or not this is due to the excessive trauma of running and vigorous games is not known. Many of these bursae are asymptomatic and require no treatment. If, however, they are tense and painful and interfere with prosthetic wearing, they should be removed. Aspiration, firm binding, crutch-walking and instillation of cortisone have all been tried, alone and in combination; they produce only temporary alleviation of symptoms. Complete surgical excision is curative and seldom requires the sacrifice of the bony stump.

Neuromata are the other cause for surgical revision of amputation stumps. They *are very infrequent in children.* In 196 post-surgical amputees, only 3.2 per cent of cases needed revision for neuromata. Surgical excision of the painful neuroma is the treatment of choice. To bury the nerve deep in stump musculature is desirable. Injection of the nerve is not recommended.

**Conversion of a Deformity to an Amputation.** *The appendicular* abnormalities that may be treated as amputations may demand considerable exercise of mature

judgment as to whether such patients may be fitted with nonstandard prostheses primarily or conversion made surgically to a more satisfactory stump and standard prostheses fitted.

Each type of surgical procedure presents its own problems. It is not our purpose here to review amputation surgery. The technics are well covered elsewhere. Most of the technics of adult amputation surgery are applicable to amputations in children. However, it will be well to review some of the differences in children.

When doing an amputation on an adult under the stress of an emergency situation, one has to plan on finishing with an adequate stump that will be comfortable and capable of being fitted with a prosthesis. In children, this desideratum may be modified, in that the stump may be one temporarily satisfactory but may be converted to a more ideal stump later. At first consideration this would seem to be a minor difference, but it is very important. Since a child is undergoing symmetric, proportionate longitudinal growth of the extremities, the operating surgeon must take growth potential into account

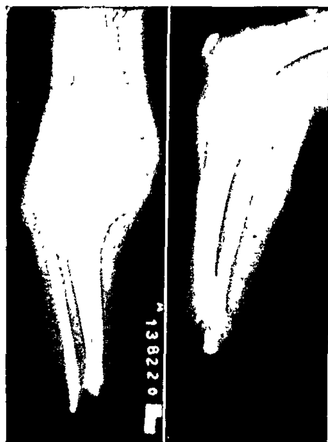



*Fig. 2. Fibular overgrowth in the bony stump of a 7-year-old boy.*

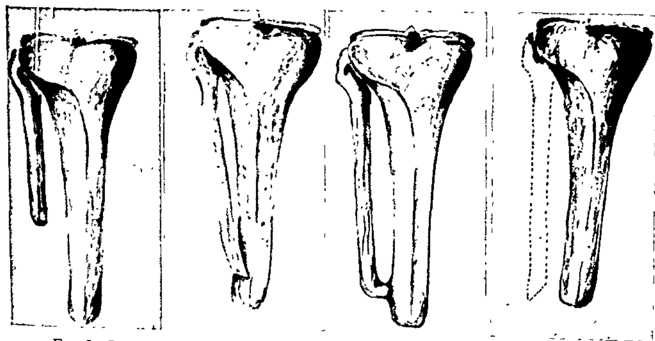


Fig 3 Surgical treatment of overgrown fibula (*From left*) High resection Wedge synostosis (after D. McKeever, personal communication). Angular osteotomy (redrawn after Barber: J Bone & Joint Surg 26:356-362). Total extirpation.

when determining the level of amputation. For instance, an ideal mid-thigh at age 5 will be a too-short above-knee at maturity; a too-short below-knee at age 5 may be an adequate below-knee at maturity, due to proximal tibial epiphyseal contribution. These point up the necessity of planning amputations in children on the basis of what they will be at maturity, not what they are at the moment. Disarticulations are preferable in children. All length possible should be preserved. Although skin grafts are not desirable, they can be used in children to save joints and length. They may require later revision.

## CONVERSION OF EXTREMITY ABNORMALITIES

The surgery for conversion of an extremity abnormality to an amputation is a complicated subject. In recommending such a procedure it is necessary to have available all the facilities to carry out the postsurgical rehabilitation of the patient. One must have sufficient experience in standard amputations to be able to predict what can be expected from conversion to amputation.

Many of the congenital appendicular abnormalities considered for conversion are total limb problems. There are subnormal musculature and joint mobility in the proximal joints. These are serious considerations, and they must be evaluated carefully before surgery is done (Fig. 4).

Not all deformities are best treated by conversion to amputation. Each case must be evaluated on the basis of the knowledge, the judgment and the experience of the operating surgeon, including a thorough acquaintance with prosthetics. The decision to convert must take into account the training facilities available to complete the rehabilitation procedure (Figs. 5 & 6).

The families of children with bizarre or abnormal extremities are understandably distraught and quite desperate for help. They need a clear-cut, detailed explanation of the total plan of treatment. The demonstration of the results in similarly handicapped children is often very helpful.

From the mechanistic standpoint (prosthetic restoration) the juvenile amputee is defined as a patient 14 years of age or younger with an amputation, either traumatic (postsurgical) or congenital. Many cases come within this definition with extremity abnormalities and may be treated by prostheses with or without surgical conversion to amputations.



FIG 4 Anomaly of right hand and forearm Limited elbow motion. The lack of development of fingers and metacarpals precludes attempts at phalangization Disarticulation at wrist offers nothing; therefore, fitting with a voluntary-opening device.



FIGS. 5 and 6, same patient. FIG 5. (*Top*) Six-year-old girl with 1 digit, 1 forearm bone and 25° motion at elbow. Paraxial ulnar hemimelia. (*Bottom*) Attempts at primary prosthetic fitting with voluntary-opening device; limited function resulted. The family was dissatisfied with the function offered, due to limited elbow motion and the unsightliness of the anomaly, and requested surgery. (Top 3 illustrations from left—Frantz, C. H.: M. Times 87:624-625)

## THE AGE OF FITTING

Determination of age of fitting presents many problems. If the patient is a lower-extremity amputee, he should be fitted as soon as he manifests a desire to stand and walk. Usually there is no problem in convincing the family that early prosthetic fitting is desirable (Fig. 7) Both family and patient readily accept a lower-extremity prosthesis. The young amputee will learn to utilize the limb quite readily and, generally, continues to remain a good wearer. The problems of cosmesis versus function do not arise. There is seemingly less rejection of a child with an artificial leg than of one with an artificial arm.

There is a divergence of opinion concerning the age of fitting in upper-extremity cases. A sound basic rule is to fit an appliance about 1 year prior to formal schooling.

At 4 years, the motor-kinesthetic pattern of the patient has developed to a point at which he has purposeful grasp and release; he has the ability to pronate and supinate, and his environmental activities are requiring some two-handed functional patterns If a patient is fitted at 4 years of age, he is sufficiently mature to benefit from training, and his limb rapidly becomes a functional attribute. By the time he is admitted to school, a year later, there is good prosthetic acceptance, improved function and an awareness of the potentials of the limb. Further, the newness of the limb has worn off so that he is not abnormally sensitive about his appliance and can often justify it by his improved function (Fig. 8).

Recently, very early fitting of upper-extremity cases has been tried. MacDonell[11] reported 12 such cases. There are sound academic reasons for doing this. Observation of these and similar cases would indicate that prosthetic tolerance is easily developed (Fig. 9). It is not yet clear whether or not those fitted very young learn how to use a cable-controlled prehension device more readily than those fitted at a later date.

The families of children who are fitted very young are gratified. They seem to appreciate having something done when they are most anxious. This is advantageous, as to obtain and maintain familial rapport and co-operation enhances the chance of successful prosthetic rehabilitation in the juvenile amputee.

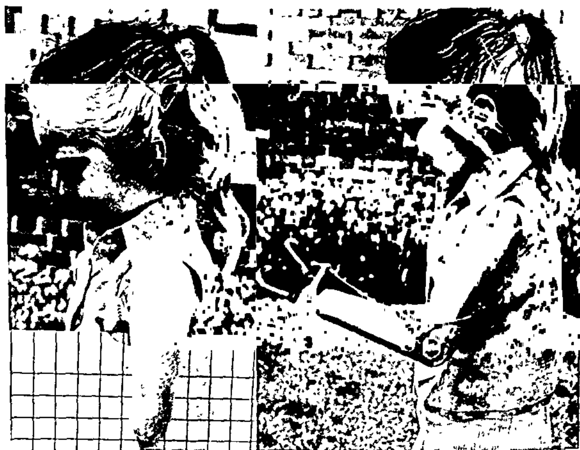


FIG. 6 Disarticulation at the elbow, preserving distal humeral epiphysis Fitted with elbow-disarticulation type of prosthesis with outside elbow lock The parents and the child were happier with functional results after guided inpatient instruction in the operation of the elbow lock and terminal device in dual combination. (Illustration at left—Frantz, C H M Times 87:624-625)

FIG. 7. Congenital anomaly of the lower extremity with extreme shortening. As soon as the baby stood up in his crib, he was fitted with a simple pylon device and the toddler's harness. Note terminal crutch tip. This is superior to the conventional caliper bar. The crutch tip allows nonskid and pivoting in gait and turning.

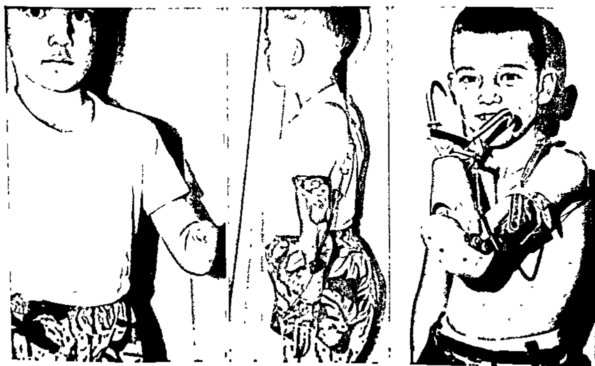


FIG 8. Congenital very short below-elbow amputation (partial hemimelia) fitted with voluntary-opening terminal device at 4 years of age, with 1 year of wearing before entering school. Very acceptable performance.



FIG 9. *Infant, 6* months old, fitted with passive mitten (acheiria, absence of the hand).

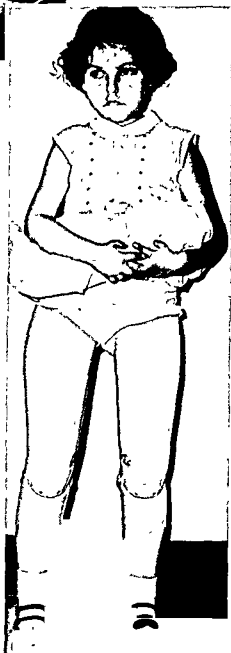


FIG. 10 This child was fitted with bilat eral suction sockets.

## PROSTHESIS PRESCRIPTION

Prostheses as prescribed currently are a series of selective, efficient mechanical components to be assembled and fitted to the patient by means of a comfortable harness and some type of suspension and control. The problems in the case of the lower extremity vary greatly from those of the upper extremity.

*It is impractical to present a sample pre*scription and limb for each type of case. Variations in component preference and fabrication technics require an atlas type of presentation. Therefore, it seems expedient to present some of the more usual prescription problems and to clarify them.

### LOWER EXTREMITY

**Above Knee.** Suction sockets work very well in children. With few exceptions, it has been the authors' practice not to apply them in children below the age of 10 (Fig. 10). Younger children are unable to apply the prostheses without help. Often this creates problems at school or away from home. Certainly they do not seem to be desirable in children who have not been toilet-trained.

Theoretically, variable friction knee joints are desirable for all above-knee cases. In practice, they are not necessary for children.

FIG. 11. Patient with bilateral congenital knee disarticulation. This child is being fitted with bilateral suction sockets. The adjustable leg is utilized to facilitate alignment and gait analysis.

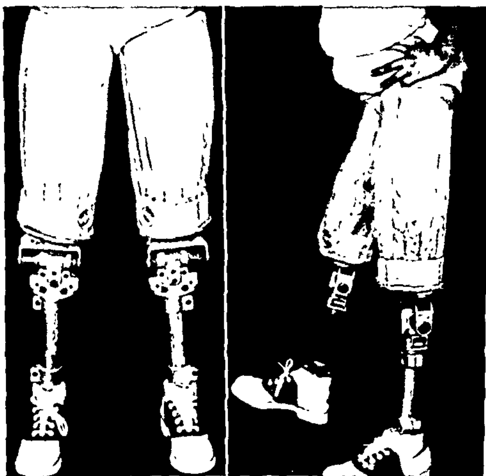


FIG. 12 Two-year-old child, congenital below-knee amputations (bilateral lower, partial hemimelia). The toddler's harness is employed to stabilize the prostheses.

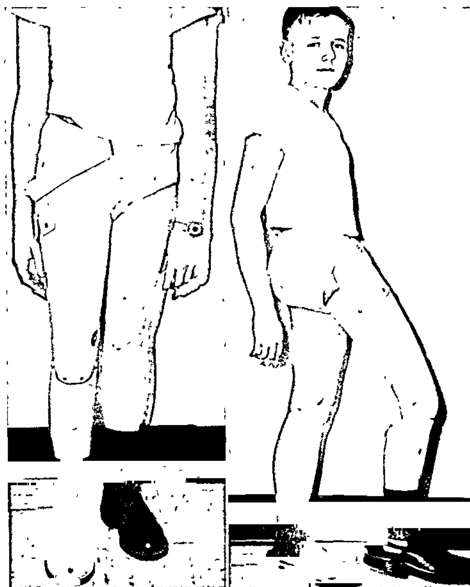


FIG. 13. Above-knee coverted congenital amputee, with dysplastic hip. Note the Silesian bandage in the trochanteric area to stabilize the prosthesis.

The gait of most children is a constant variable, and training attempts to make it ideal are impractical. This adjunct (variable cadence knee) to good prosthetic rehabilitation is not necessary in children.

Fit and alignment are mandatory at all ages. When size permits, the use of an adjustable leg is helpful (Fig 11). Efficiency and conservation of energy are facilitated by good fit and alignment.

Suspension other than suction of above-knee limbs will depend entirely on the patient's age In the very young, the "toddler's harness" has been very helpful Stability, as well as wider distribution of prosthetic weight, is increased (Fig 12).

Standard hip joints and pelvic belts should be reserved for older patients. Silesian bandages or web-elastic modifications of them seem to be preferable in the presuction cases older than the toddler group (Fig. 13).

When fitting the very young above-knee amputee with his initial limb, it is believed to be desirable to start with the knee fixed in full extension (Fig. 14). This permits more rapid development of alternating progression. Once balance and confidence have been established, the knee may be unlocked gradually, and finally a full-swinging knee is obtained.

### UPPER EXTREMITY

Here the problem is quite different from that in the lower extremity. In the lower

extremity, the major functional replacement required of a prosthesis is weight-bearing. This may be articulated or not, and may have multiple refinements.

In the upper extremity, a multitude of functions have been lost. These include mobility, prehension of a wide variety, tactile sensory function, strength, agility, cosmesis and varying degrees of skill. No prosthesis can replace all these lost functions. The prescription-writing team must determine which functions can be replaced and prescribe prostheses that will restore them as efficiently as possible. There are choices relative to size, shape, strength and weight. The problem is to determine which of the various components available are most desirable for the patient in question. The answers require a wide knowledge of the components commercially available, including the advantages of each, plus a sufficient knowledge of the patient's potential to permit the matching of components to functional needs in a manner that will develop adequate (desirable) function and ensure amputee acceptance.

Terminal Device. This is probably the most critical component in upper-extremity prostheses. These devices range from the hand with cosmetic glove to the simplest



FIG. 14. A 3-year-old congenital quadruple amputee; bilateral above knee. Note the anterior locking straps to prevent buckling on initial ambulation.

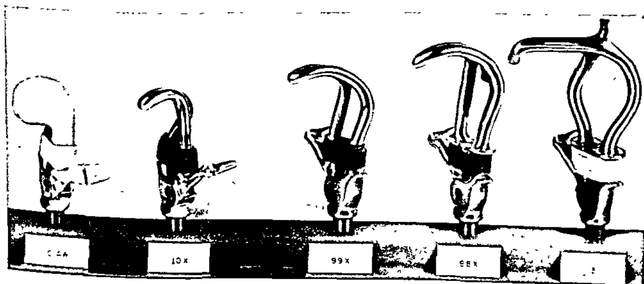


FIG. 15 Conventional types of voluntary-opening devices Extreme left: the plastic-dipped wafer hook for toddlers.

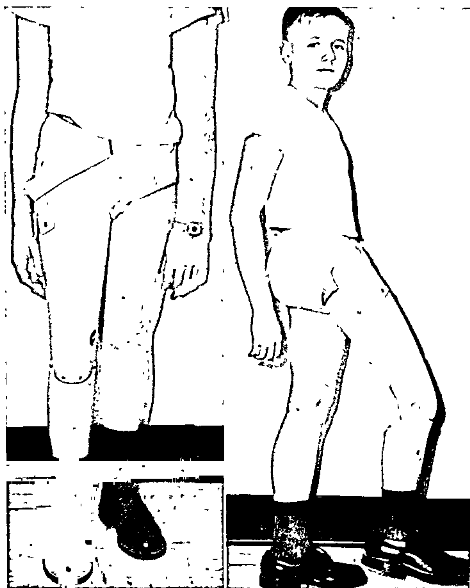


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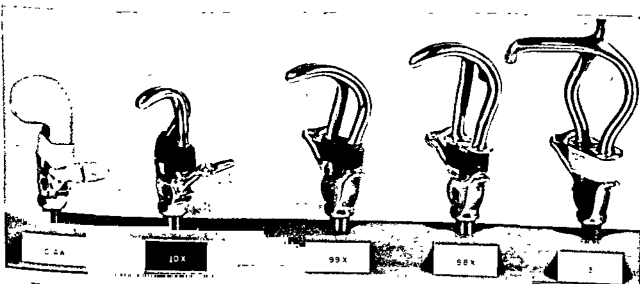


FIG. 15. Conventional types of voluntary-opening devices. Extreme left: the plastic-dipped wafer hook for toddlers.

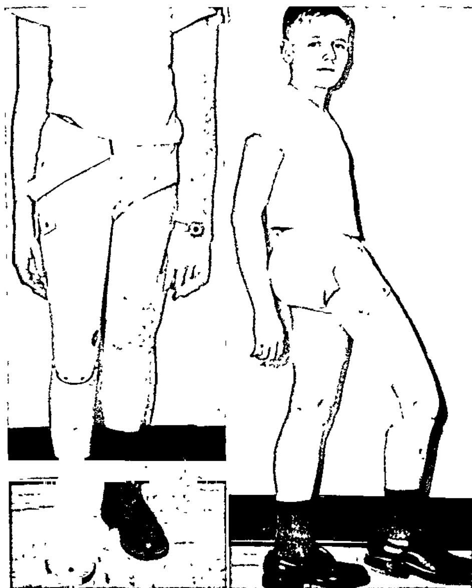


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FIG. 17. (*Left*) Classic phocomelia, lower extremity. (*Center*) Fitted with plastic pelvic mold with a child-sized Canadian hip disarticulation unit. (*Right*) Lateral projection demonstrates axis of the joint relative to body alignment.

the infrequent use of more than one terminal device. Wrist flexion units are not generally available commercially, and, except in the severely handicapped (bilateral upper-extremity amputees), they have little to add functionally.

**Elbow Units.** There is a multiplicity of below-elbow hinges. There are reasons to justify most of these variations. The prosthetist's preference seems to be an excellent way of determining which type will be used.

The variable ratio hinge is desirable in the split-socket fitting for the very short below-elbow amputee. The standard step-up ratio is 1:1.5.

Most above-elbow units are basically cable-controlled multiposition elbow locks. Some are an enclosed unit; others are exposed units. The exposed ones are necessary in elbow disarticulations to prevent abnormal length of the above-elbow segment. In all other cases the enclosed type is preferable. There are other mechanical variations on this basic pattern that may be used. Here, again, the determinant primarily is familiarity with and confidence in the mechanical component.

**Cables.** The metal Bowdin type of cable is now generally considered to be standard equipment. The nylon-lined cable housing has done much to reduce noise and to improve efficiency. Leather or nylon lanyards seldom are indicated.

**Harness.** Dacron tape is a suitable dimensionally stable material. Its width should be varied according to the patient's size.

The pattern is variable. Unquestionably, the figure of eight is the basis of all upper-extremity harness patterns. Additional suspension is desirable at times and is to be used as needed. Most skilled prosthetists become adept at varying harness patterns to the individual. All members of the prosthetic team must be familiar with harnessing and its variations. In the upper-extremity case, the harness both suspends the prosthesis and is the mechanism for the transmission of power. These are critical func-



FIG 16. The voluntary-closing APRL (Army Prosthetics Research Laboratory) type of terminal device. Interchangeable with APRL type of voluntary-closing hand covered with plastic glove offering cosmetic restoration.



two-fingered voluntary-opening hook. Units are made in steel and aluminum. Finger shapes vary according to work requirements (Fig. 15). They may either open voluntarily or close voluntarily. Some are spring loaded, some rubber-band loaded, and those that close voluntarily are of variable loadings, depending upon the input through the cable. The hands vary from the purely cosmetic (nonfunctional) restoration to the voluntary-closing functional (APRL) hand.

The selection is exceedingly difficult. Many factors must be weighed. Age and size are probably the two main determinants in children. Age determines the skill requirements and whether light-duty or heavy-duty equipment is indicated. Size narrows the field of selection, because it is primarily in the voluntary-opening type of devices that there is a sufficiently wide variation in size to permit the proper fitting of young children.

There is no rule of thumb. It has been the authors' practice to fit a passive mitten under 24 to 30 months of age (see Fig. 9). From this level to the teen-agers we have used a voluntary-opening two-fingered hook of adequate size, interchanging steel or aluminum on the basis of the child's functional needs (see Fig 8). At the teen-age level, the increasing social awareness of the patient requires some concession to cosmesis, so the voluntary-closing (APRL) hand often is ordered (Fig. 16). There have been exceptions to this practice. It is believed that this is an adequate baseline on which to develop experience and from this later to initiate discriminatory, variable terminal devices.

At the present stage of development, there is no doubt that the two-fingered voluntary-opening hook offers the greatest functional restoration. There is a minimum of upkeep, and the cost is reasonable. However, this device is the one least desired by the parents. Because of its many advantages it is worth while to take time enough to promote acceptance of the voluntary-opening hook. Once accepted, its intrinsic functional worth will be acceptable to the young amputee and his parents.

**Wrist Units.** In children there is infrequent need of anything but the simple friction-type device. The F. M. (Fletcher-Motis) disconnect is seldom necessary because of



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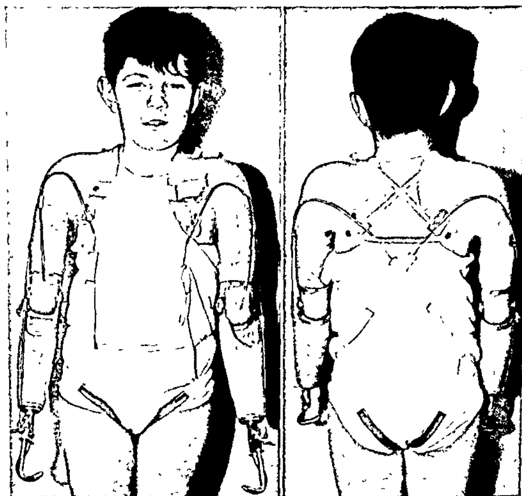
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tions. All good upper-extremity prostheses depend upon expert harnessing for good function.

In this discussion there has been avoidance of the hemipelvectomy and hip-disarticulation cases as well as the shoulder-disarticulation cases. These 3 amputation types, although statistically infrequent in children, represent some of the most difficult cases. Each is an individual problem and requires a kind of custom evaluation and fitting. The Canadian type of hip-disarticulation prosthesis is certainly an advance in fitting this type of case. It can be fabricated readily for children and has proved to be desirable (Fig. 17).

At present, shoulder disarticulations are fitted with rigid shoulders (no humeral abduction, flexion, extension). This is less than desirable, but there are no items commercially available that permit shoulder motion. Experimental work is progressing, and conceivably there may develop a unit for shoulder disarticulations that will permit limited stable shoulder motion (Figs. 18 & 19).

In the hemipelvectomy cases, the major



FIGS. 18 and 19, same patient FIG. 18 Conventional type of shoulder disarticulation prostheses Note that the shoulder cap and humeral section are one piece, allowing only forearm flexion and elbow action in the frontal plane. Elbow-locking mechanism controlled by waistband, and terminal device operation controlled by perineal strap No rotation or abduction is possible, except by prepositioning of the turntable incorporated in the elbow joint.

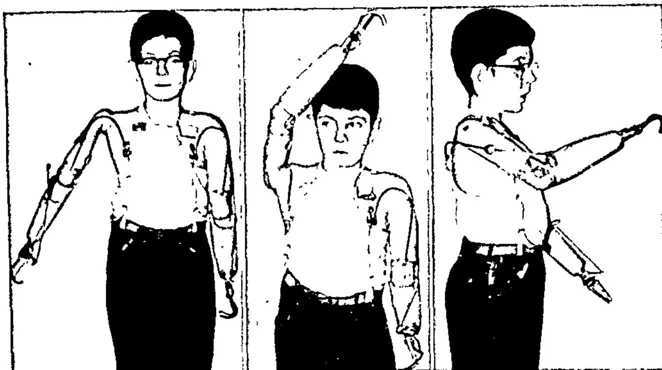


FIG. 19. Experimental type of ball-and-socket prepositioning friction shoulder joint. Prepositioning requires an assistant or excessive body motion, but range of operation is improved.

requisite is the fitting of a comfortable, stable torsopelvic socket. This requires custom prosthetic work and critical attention to details of fit.

A good juvenile amputee service requires a co-ordinated team to evaluate, prescribe for, fit and train the patient. This necessitates a detailed knowledge of the problems and the requirements of the juvenile amputee, be he a postsurgical amputee or a true congenital amputee, or the patient with an abnormality who is being treated with a prosthesis.

Clear recognition of how the child differs from the adult is mandatory. This is true both at the time of amputation/conversion and at the time of prosthetic prescription-writing and training.

Inpatient training services are desirable. Experience has shown that such services, though costly initially, produce greater prosthetic acceptance and, thus, more wearers and better rehabilitation.

Careful follow-up is also a necessity. Each patient should be seen at least every 4 months. Patients do not wear mechanically unsound or uncomfortable prostheses. In many instances the rigors of childhood will produce mechanical failures as often as every 4 months. The progressive longitudinal and circumferential growth of the preadolescent will require such frequent observations to maintain fit and alignment.

Conservatism is urged. Fit the child, train him to use the prosthesis, then observe him. Surgical revision (conversion) can be interjected at any time. Experience will demonstrate that it is required less often than one would suppose. Most children, comfortably fitted with mechanically sound prostheses, will function very well. These patients have a rehabilitation potential that is generally greater than the victims of severe postpoliomyelitis and cerebral palsy, and they deserve diligent care.

## RÉSUMÉ

The ideal program for rehabilitating the child amputee must be well organized and

staffed with a group of specialists who comprise a team (see Fig. 1).

Experience over a 12-year period has shown that parental participation and co-operation are important factors that may spell success or failure.

The social service worker initiates the program. It has been found that a pre-physical orientation conference will acquaint the patient and the parents with the staff and the routine to be expected, and answer many questions relative to time, travel and correspondence. It also establishes rapport. An insight is obtained relative to the family's reaction to their child's problem. Further, they are assured of the interest and the sympathy of the medical staff.

The examination of the child and the amputation stump is expedited in the clinic. Here the necessity for physical therapy is determined. The newly amputated child may need preprosthetic conditioning of skin and muscle-building exercises. Prosthetic fabrication and fitting may be initiated immediately or may be delayed until stump conditioning is judged to be satisfactory.

Prosthetic training is begun when the new prosthesis is determined to be satisfactory for comfort, fit and alignment by the clinic team. This phase is pursued while the child is an inpatient. Both physical therapists and occupational therapists participate. The usual practice, when both types of specialists are available, is to direct the major responsibility to the occupational therapist for the upper-extremity amputee and to the physical therapist for the lower-extremity amputee. This does not mean that these departments are segregated, they work in close co-operation. It is their responsibility to bring the child up to an acceptable level of performance.

While inpatient training goes on, the child may attend school in the appropriate grade. Social and recreational activities may be directed to prosthetic activity, such as sports and social events where the child plays games and eats with other children, thus decreasing self-consciousness.

When the training of the child is considered to be adequate by the clinic team, the young patient is discharged. A conference is held again with the parents. The prosthesis is demonstrated in detail; its care and operation are explained. Preparations are made for going back to home and school. The parents are encouraged to work with the child and endeavor to promote increased skill with the prosthesis. This is very important in the upper-extremity amputee.

There are two critical phases for the young amputee: when the young congenital amputee enters kindergarten with a prosthesis and when the young postsurgical amputee returns to school different from what he formerly was, i.e., with an appliance. The teacher has been said to be the "second mother" for the elementary-school child. As such, she must be a member of the rehabilitation team. She should be forewarned and briefed by the parents. She will develop insight into the child and aid him in taking his place in the class group. She can prepare the class for this young amputee, have him demonstrate his skill with the positive emphasis on "how much he can do." Children are curious, and, once their curiosity is satisfied, no further questions are asked. At times, playmates are unthinkingly frank, but not cruel. The term *Captain Hook* has become a nickname for upper-extremity amputees This should not be thought of as a derogatory remark any more than *Red* for a red-haired child or *Shorty* for the youngster of short stature. Total rehabilitation is the goal of the child amputee. This implies the acceptance of the handicap by the child and his parents If properly handled, slowly but surely he will become integrated into the social, the physical and the intellectual facets of his school. The intelligent teacher who is aware of the functional capacity of the child and his limitations will aid him greatly.

Outpatient follow-up is the maintenance

portion of the program. A minimum of 4 clinic visits a year is standard practice in the majority of cases. Many cases demand more than 4 visits a year; some have from 7 to 10 visits. The clinic visit offers the opportunity to keep the prosthesis mechanically sound, modify the sockets for growth and refit new mechanical components.

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## Le Tutela del Amputato Juvenil

*Summario in Interlingua*


Recente progressos in le technicas prosthetic e nove cognoscentias concernente le amputato pediatric permitte, super le base de un ben-organisate programma, equipar e rehabilitar practicamente omne typos de amputato traumatic e congenite.

Le crescente mechanisation del mundo moderne expone le juvenes a multe episodios traumatic que resulta subsequentemente in amputationes. Assi nos ha un alte proportion de non-congenite amputatos juvenil qui constitue gruppos traumatic con respecto al etiologia de lor casos.

Le efficace tutela e training del amputato juvenil require un ben-organisate equipa consistente del chirurgo orthopedic, del therapeuta physic, del therapeuta occupational, de un qualificate prosthetista, e de un consulente social con qualificationes medical. Le consilio de psychologos, psychiatros, e pediatros debe etiam esser disponibile. Le patientes debe esser equipate con ben-construite, confortabile, e adequatemente harnesate apparatos pro functionar satisfacentemente.

Le tutela del amputato juvenil require que on recognosce le area in que le juvene differe ab le adulto. Le juvene es membro de un gruppo familial, e como tal ille depende, pro un grande parte de su disveloppamento, de su familia. On non pote expectar que le juvene se desira un prosthese in plen comprension del resultante avantages functional. Frequentemente il es le familia que debe facer le decision e que debe, subsequentemente, ager como un gruppo cooperante con le equipa professional al clinica amputatori.

Le juvenes differe ab le adultos in tanto

que illes cresce e se disveloppa in multe aspectos de lor existentia. Lor systema motoskeletic deveni progressivemente plus habile e plus grande. Isto resulta in le necessitate de un frequente adjustation del prostheses. Le fluctuationes emotional del juvenes require consiliage e le exercitio de disciplina del parte del familia.

Le casos del amputatos juvenil es classificabile in tres gruppos: (1) Casos non-congenite (post-chirurgic e post-traumatic); (2) casos strictemente congenite; e (3) casos de anormalitates appendicular.

Le problemas chirurgic del amputation pediatric se rangia in duo classes principal.

1. Amputationes de urgentia, ubi le factores etiologic es traumatisationes acute, i.e. per exemplo explosiones, accidentes vehicular, accidentes per machinas-instrumento, accidentes de ferrovia, e accidentes de ferma. Multe attention e bon judicio debe esser usate quando on se trova confrontate con le problema de un amputation potential in un juvene. Si le superviventia del extremitate in question es non ancora decidite—positive- o negativemente — omne effortio debe esser facite pro salvar lo Un amputation tardive es a preferer a un amputation innecessari.

2. Amputationes elective, ubi un major gruppo de casos es representate per extremitates non-functional como resultato de lesiones neurogene, de gangrena post fractura, de lesiones thermic, e de tumores Tal casos pote esser tractate super le base de plus extense deliberationes

In association con le amputationes elective on pote mentionar le problema del retractamento de ancian truncones. In le majoritate del casos juvenil iste problema non occurre. Juvenes non suffre le complication que usualmente plaga adultos, i e. membro phantoma, cicatrices, neuromas, alterationes de pelle trophic, e truncones troppo curte Hypercrescentia es un problema que se incontra in amputatos juvenil con operationes effectuate ante le etate de nove annos. In iste phenomeno il occurre un disproportion del crescentia in le truncon de osso e in le revestimento de histos molle. In tal casos, un revision chirurgic es indicate.

Un altere sub-gruppo de casos pone le problema del conversion de anormalitates appendicular in truncones semi-classic per medio de methodos chirurgic. Omne tal caso debe esser considerate individualmente. Le chirurgo qui se trova confrontate con un problema de iste genere debe prender in consideration le contribution de epiphyses de ossos longe. Ille debe planar su tractamento si ben e si detaliatemente que ille es capace a explicar al parentes omne aspectos del objectivo que es a attinger. Isto presuppone le disponibilitate de facilitates pro le satisfacente adjustation prosthetic e pro le efficace training in le uso del apparato.

Le etate al qual le amputato juvenil es equipate con un prosthese debe esser dictate per su habilitate de servir se de illo. Assi, quando il se tracta de un extremitate inferior, si tosto que le juvene pote tener se in position erecte, le truncon del absente membro debe reciper un prosthese, e non importa si le prescription debe ancora esser satis primitive. Quando il se tracta de un extremitate superior, le adjustation del prosthese es a effectuar un anno ante que le patiente entra in le jardin del infantia. Juvenes de quatro annos de etate tolera e activa multo ben un prosthese de extremitate superior. Si le prosthese es prescribite a iste etate, le patiente ha un anno de practica ante su entrata in un schola formal, e ille va non sentir se inferior a su companiones de classe. Le autores ha executate un experimento clinic in que infantes de etates usque al minimo de cinque menses esseva equipate de apparatos passive. Tolerantia del prosthese esseva effectuate sin difficultate, e il pare que un forma primitive de ambidexteritate se disveloppa con le dispositivo terminal del apparato al mesme distantia ab le corpore como le mano intacte. Isto corresponde a un modo fundamental de comportamento e incoragia le patiente a ingagiar se in activitates distante ab le corpore. Un del effectos

de iste methodo esseva reducer le necessitate de restringer le extremitate normal in su distantiation ab le corpore quando illo assiste le latere amputate. Il ha essite constatate que juvenes jam al etate de vinti-quatro menses pote servir se de un apparato con dispositivo terminal que es aperibile-claudibile a voluntate. A trenta menses de etate illes pote maestrar le cubito a fixation positive in varie positiones.

# 5

# Recumbency Versus Nonrecumbency Treatment of Legg-Perthes Disease

CHARLES W. GOFF, M.D.*

Conant remarked that "a theory is never overthrown by contradictory facts; it is only overthrown by a better theory." For many years I have believed in the theory of long-time recumbency care of Legg-Perthes disease (Legg-Calvé-Perthes syndrome). Many facts were marshaled that seemingly recommended this form of care. In spite of this experience, an apparently equally successful form of nonrecumbency treatment, consisting of partial or total weight-bearing from the beginning, has been practiced by a reputable group of orthopaedists. Their results seem nearly to equal those following my recumbency method. How could this dilemma be resolved?

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The adequacy of any theory should be judged by certain criteria. In this instance: (1) Are the facts known about the disease accounted for? (2) Could new observations prove to be profitable? (3) Would these observations be related to yet another set of facts? Accordingly, I have proposed *a new theoretic model* based on fresh facts, indicated by recent mechanical, circulatory, microbiologic and postural information. These relationships had been obvious for

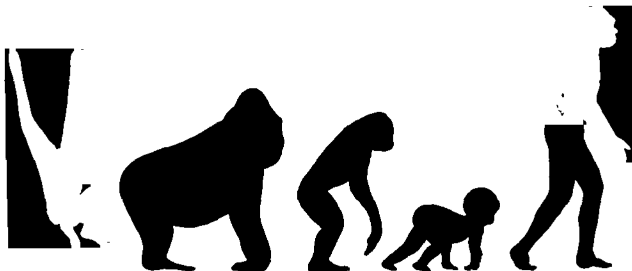


FIG 1 Erect stance of primates culminated in man's forward rolling pelvis and nearly complete extension of the femur at the hip joint. In this way full, bipedal weight-bearing evolved The center of gravity was pulled posteriorly—greater stress and strain was placed on the head of the femur From left to right are depicted the brachiating gibbon, the gorilla, the chimpanzee, a man child and an adult.



Fig. 2. Todd gorilla, showing coxa plana of left femur following Legg-Perthes disease (?). Lower views illustrate the area of weight-bearing, larger than in man, and preserved quite well The associated acetabulum, not reproduced, shows good spheroidal molding Animal lived to about 18 years of age, when he was shot by a hunter. The gorilla must have been handicapped, probably had pain and limited locomotor capacities (Taylor, King & Stecher: Osteoarthritis of the hip in gorillas *in* Clinical Orthopaedics No. 6, pp. 149-157, Philadelphia, Lippincott)

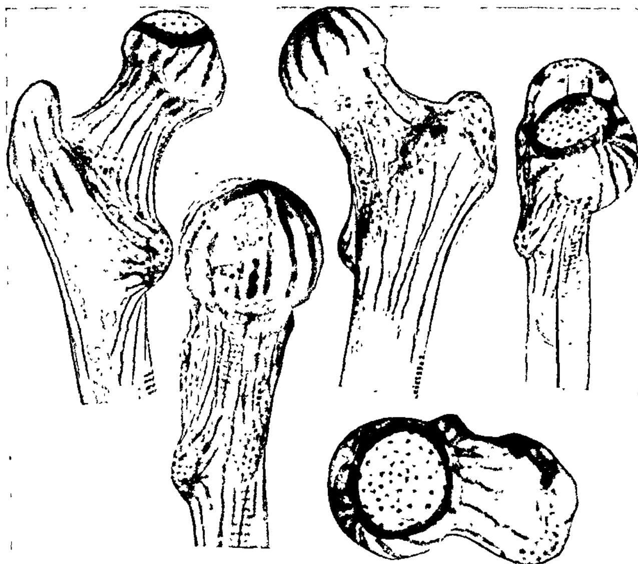


FIG 3. Upper femur of man Split lines and stresscoat markings outlined in India ink illustrate locations and dense subchondral bone of the weight-bearing surface. Note the posterior medial geographic site of this zone. Trabeculations from this plate converge on the medial neck and the upper shaft cortex.

some time to a number of investigators. My new theory has restricted prolonged recumbency to those children whose capital femoral epiphysis and metaphysis showed an involvement of the weight-bearing area. If this region was spared, or demonstrated rapid recovery, weight-bearing could be tolerated at an early date This also would aid in restoring normal physiologic responses, and an early recovery with better globular femoral heads should be rewarding. Furthermore, such an alternate theory of selective weight-bearing may explain some of the good results obtained by those orthopaedists who did not prefer recumbency for these children. My dilemma then would be resolved What follows is not an apology, but an apostasy—a new article of faith.

## POSTULATES OF THE PRESENT THEORY

Man's hip joint, as a part of his pelvis, has retained many features of a quadruped. An infant's hip does not extend fully during the first 24 months. A child learns to walk with a bent knee and a partially flexed thigh,

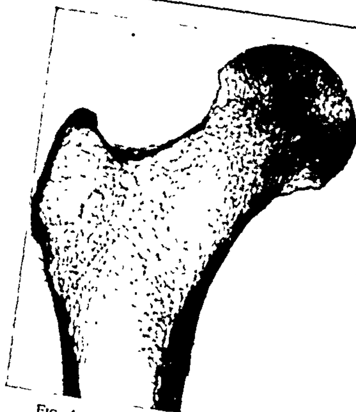


FIG. 4. Upper femur of man. Photograph by transmitted light of a thin section indicating trabeculae of support. The femoral calcar is not shown. This is located in another plane.

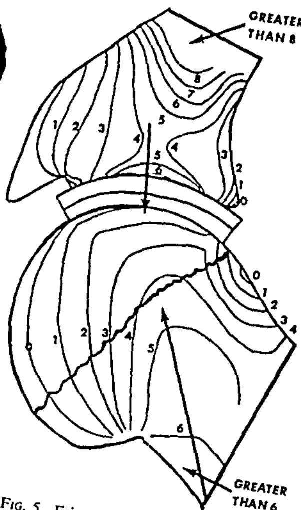



FIG. 5. Fringe patterns of shear strain. The hip joint, as reported by Fessler; numbers indicate values that he obtained. Wavy line is the epiphyseal plate drawn in, and the arrows indicate weight-bearing direction in erect standing position. Note the small zone of greatest strain. (Redrawn from Fessler, H.: J. Bone & Joint Surg 39-B:145-154)

as does a young chimpanzee. The latter never acquires full extension of either joint, but the child may. The pelvis has been recognized as an evolving unit of body support not yet fully adapted to the erect position. There persist a number of deficiencies that lead to hip and low-back disorders. The great apes—chimpanzee, gorilla and orangutan—appear to be free of these disturbances to a greater extent, although Taylor, King and Stecher[20] have reported an occasional occurrence in these interesting primates.

**Adaptations of Posture.** Man's femur becomes extendable to 180° with his body; accordingly, the erect weight-bearing femoral surface has become smaller and displaced more posteriorly. Man's globular femoral head has reacted according to the laws of fluid dynamics and opposing frictional surfaces, locking tightly within the acetabulum in the extended position.[1] Other primates do not possess this capacity. Such a mechanism is found in all varieties of Homo sapiens. Trauma to these structures occurs more easily,[8] and the consequences are greater. The "adaptation syndrome" raises its ugly head, even in children.

Locomotor studies of all the primates indicate a unique pattern of weight-bearing in man. The femur swings through an arc of about 35° when a normal step is taken, either running or walking.[1] Illustrations show a secondary expanded elliptical weight-bearing zone of the femoral head, the anterior portion of which bears weight at the beginning of the propulsive phase of loco-



Fig 6 Circulation of upper femur of child. Illustrates the greatest supply from lateral and posterior sources rendering the medio-anterior area more labile to vascular damage. (Trueta, J.: J. Bone & Joint Surg. 39-B:358-394)

motion in man; the posterior part supports the body weight when standing completely erect. Cross sections indicate a more dense cortical bone under the articular cartilage of this area, pointing to osteons that seem to terminate in this region. Split-line and stresscoat patterns[3] have determined this zone This also is true at trochanters, where the bone is relatively thin but shows a pattern of terminal osteons rather than osteons arranged as in the outer cortex of the femur An observer seems to be looking down on the open end of columns of osteons rather than at their outer surfaces. This pattern correlates with the trabecular concept of many students of bone mechanics from early days to the present.[2,5,10,15,18] Actually, the trajectorial arrangement represents a combination of growth directives inherent within each species, oriented according to a specific bony shape and influenced by adaptability to environmental stress and strain [3]

**Blood Supply of Upper Femur.** Arterial blood enters the femoral head, neck and upper shaft through three sources. The most important is the nutrient artery, running a tortuous course through the cortex of the shaft to reach the intramedullary cavity and bone marrow. Within this cavity it divides and sends branches upward and downward toward the metaphyses of the femur. At intervals small branches are given off that pass to the inner surface of the cancellous bone through horizontal Volkmann's canals. These anastomose extensively with small arterioles that enter the bone from the periosteum and with extensive venules from all sources.

The epiphysis of the upper femur is supplied by two arterial systems, the posterior half of the epiphysis being supplied more generously than the anterior portion. The former contains the geographic weight-bearing center.

Many circulatory studies also indicate a variety of arterial and venous patterns about the proximal femur,[19,21,24] depending on age and genetic factors. The clearest are those

of Trueta[21] and Tucker.[21] Recently, Trueta has proposed that the latter probably is responsible for the great rarity of Legg-Perthes disease in the African and the American Negro. As I suggested in 1954,[8] there is a racial predilection in the Negro, pointing to the Middle and the North European racial stocks as the most likely gene pools from which this disorder flows.

Anatomic Relations of the Acetabulum. The articular surface is narrow, elliptical and concave. It is similar to a race of a universal shaft bearing. The ligamentum teres with its blood vessels enters from below,[11] as it does in all quadrupedal animals, and is a non-weight-bearing area. Only a small zone of the head of the femur is in contact with the acetabulum during weight-bearing.[7]

Biopsies. Jonsäter[13] biopsied many capital femoral epiphyses by inserting a punch above the cartilaginous growth disk into the cancellous bony nucleus. In the early critical phase of the disorder there are dead bony areas intermixed with fresh osteoid tissue that correlate with the dense roentgenographic patterns. Translucent regions show various stages of osteogenesis. Unpublished data by Gee indicate an above-normal blood supply existing within the

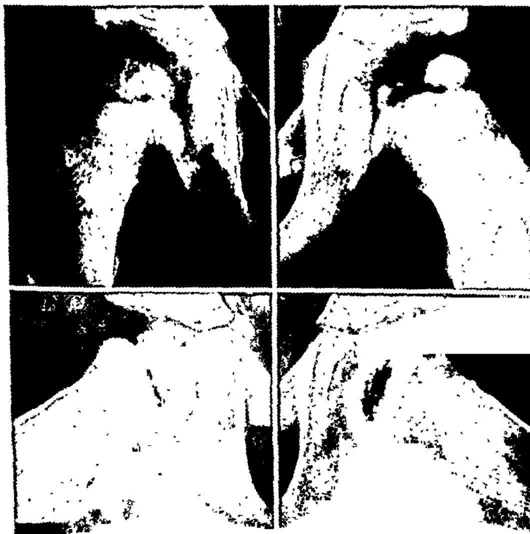


FIG. 7. Early unilateral partial Legg-Perthes disease Illustrating the trochar channel (*left*) made by Gee in his radioactive isotope studies of the circulation of zones of femoral epiphyses during the disorder The more translucent area had the greatest blood supply—was regenerating bone at a fast rate

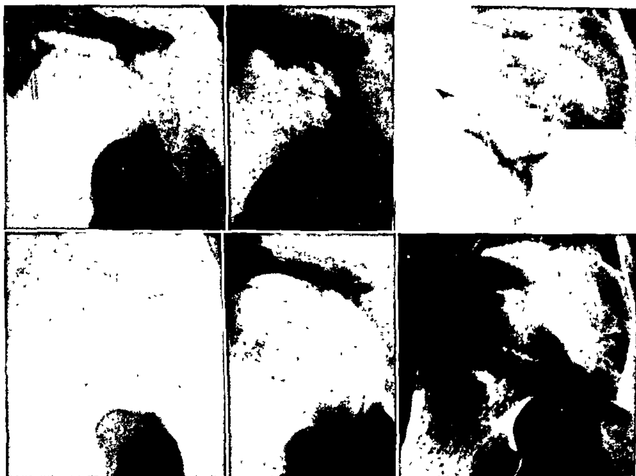


FIG. 8 Roentgenographic patterns—total epiphyseal involvement Note typical coxa magna at an early date Right upper and lower are 12-year end-result plates Poor result in spite of a long recumbency.

translucent areas of the ossific nucleus. These have been regarded as absorptive zones of the epiphysis, whereas actually they are regions of early osteogenesis. In later stages of the disorder, there is observed a demarcated region in which bone has been spared the avascular effects. This is especially true in those instances of partial epiphyseal involvement A selective effect is apparent, sparing bone in various amounts.

## CLASSIFICATION OF ROENTGENOGRAPHIC PATTERNS

For many years complete life-cycle roentgenographic studies have been maintained at Newington Hospital. Over 150 of these children, who have been treated in various ways, but chiefly by long-term recumbency, have reached a point of recovery. These were available for analysis. By inspection their early roentgenograms were divisible into three general patterns: Type A, total epiphyseal involvement (37%); Type B, bilateral epiphyseal involvement (13%); and Type C, fractional or partial epiphyseal involvement (50%). These are illustrated by accompanying life-cycle roentgenographic studies.

**Roentgenographic Patterns Correlated With Regional Circulation and Weight-Bearing Zone.** Practically always, those cases that showed a *partial epiphyseal involvement* were represented on roentgenograms by an immediate increase in density of the anterior two thirds of the epiphysis. Within 2 to 6 months, the anterior and the lateral portions of the epiphysis and the metaphysis became translucent as the circu-

Fig. 9. Partial involvement, sparing weight-bearing zone of epiphysis and metaphysis. Plates marked "W" indicate pattern that permits guarded crutch-walking. Note good results 26 months after onset.

(A) Onset age, 5 years. (B) After 8 months' recumbency. (C) Pattern after crutch-walking for 6 months' partial weight-bearing. (D) Pattern after crutch-walking for 12 month's full weight-bearing. (E) End-result after 26 months. Patient given 50 mg. of achrovim daily for 26 months.




lation rapidly returned The more dense bone—that is, the avascular bone—was rapidly absorbed, and the untouched, or living, bone returned quickly to its normal bone trabecular pattern. This pattern indicated a sparing or rapid return of the weight-bearing zone of the epiphysis. These children were allowed to walk with crutches, usually within 4 to 8 months after onset of the disorder. No deleterious results have been observed; instead, a rapid response has followed, with an earlier reossification of the remainder of the epiphysis and the metaphysis.

When the *entire*, or *total*, epiphysis \

not necessarily a poor result; on the contrary, many end-results are of this variety. Some discomfort may come on after 35 years of age, but this is rarely severe. The degree of decompensation of such a hip joint depends on the globularity of the restored femoral head and its fit within the acetabulum. If these children are treated properly, I believe that such a fit will prove to be quite adequate. Symptoms of discomfort are postponed for 25 to 30 years, and they may never occur. Only very long follow-up examination will provide valid statistics.

**Bilateral Involvement.** Trueta would classify these children according to each hip. This is a valid distinction, but for practical reckoning I have listed them separately. Since they represented only 13 per cent of our series, they will influence statistical results in a small way.

Because of the likelihood of involvement of the other hip, all children should be kept recumbent at least 4 months, during which time signs usually will appear in the other hip if it, too, is going to be affected. This second femoral epiphysis will pass through the several stages more rapidly and will be quite ready for guarded crutch-walking with full weight-bearing by the time the first hip has reossified sufficiently.

## RESULTS

This is not an end-result study. There are many in the published literature,[6,17] including one of my own,[8] all of which are of small value when comparing one series with another. In this instance, over 75 children have been allowed to bear weight at an early date. The original average length of recumbency of Pike[16] was 27 months in 29 "healed" cases; Gee and Gossling reported an average of 44 months in an additional 60 cases, many of which had not reached full recovery Katz[14] reports an average bed rest of 18.3 months for his good and 20 3 months for his fair results. The difference is not significant In my series, the time of recumbency has been reduced gradually from an average of 19 months, as reported in 1956,[9] to my present average of 9 months. When a final computation is made, this probably will be reduced even more. Those with a partial involvement are permitted to bear weight after 2 to 4 months' recumbency. To date, no deleterious effects have been observed in 45 children screened by this system.

## CRITERIA TESTED

(1) A number of facts related to Legg-Perthes disease have been verified. Other facts will become known in time and may be the means of altering my theory. (2) New observations have been examined and correlated. (3) These correlations have validated the new theory so far as they go, establishing certain conclusions.

## CONCLUSIONS

Unquestionably, immediate recumbency is beneficial. Stress and strain are relieved at once, and the child's disorder then may be evaluated properly. The selective geographic area involvement of the femoral epiphysis can be determined. If the weight-bearing region is affected, recumbency should be prolonged until this area has been restored sufficiently to withstand molding by the acetabulum under erect weight-bearing With sparing of the region, recumbency may be discontinued as soon as this fact is apparent on roentgenographic examination. Fifty per cent of my children had limited or partial involvement sparing the weight-bearing area. *For these children early crutch-walking proved to be beneficial.*

Thus, one half of the Newington series falls within the division wherein the value of recumbency was limited. Since all children were put to bed initially for a few weeks, continuous weight-bearing cannot be judged against my series of results. However, the nondeleterious effects of weight-bearing in these selected instances (50% of the series) after 2 or more months of recumbency were striking. These facts sub-

antiate in part the school of therapy using ss recumbency plus early walking care. he dilemma is partly resolved.

Total epiphyseal involvement requires uch longer recumbency.

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## Tractamento a Decubito Contra Tractamento Non-decubital in Morbo de Legg-Perthes

*Summario in Interlingua*


Es proponite e testate un nove theoria que modifica le prescription de decubito prolongate pro omne patientes pediatric con morbo de Legg-Perthes. Nunc solmente pa-

cento bilateral. Iste ultime typo require un plus prolongate decubito. Nove factos concernite con limitationes adaptational del coxa, studios special del femores de primates (inclusive investigationes de lineas de fissura), le disposition selective del vasos sanguinee, biopsias, e characteristicas roentgenologic es correlationate con le constatationes clinic.

# The Present Trend in Treatment of Osteogenic Sarcoma

ALBERT B. FERGUSON, SR., M.D.*

... session was devoted to bone tumors. Much of the discussion concerned osteogenic sarcoma ...

sought.

## RESULTS OF PROMPT AMPUTATION

Green[1] reported that all patients treated for osteogenic sarcoma at the Children's Hospital, Boston, in the period 1928 to 1946 had died in less than 27 months, with a single exception in which the diagnosis was subject to question. The treatment was prompt amputation.

Coley[2] stated in 1949 that the group under the age of 10 at the Memorial Hospital, New York, showed only 1 survival after amputation for osteogenic sarcoma, ...

... -year survivals apparently free of disease ... first ... 2 of ...

-23 in the third month and 8 of 73 in the next 3 months; thereafter results were somewhat better.

It is not credible that the deadly results of prompt amputation very early in the disease are explainable by greater malignancy in such cases. That explanation was not believed by those who studied and reported the cases in question. In reporting on 82 Registry cases seen within 2 months of onset of symptoms, Ferguson[3] noted that 17 were judged to be of unusually high malignancy. They formed 13 per cent of those having amputation within 2 months of onset, 25 per cent of those amputated at 3 to 6 months and 30 per cent of those amputated later. The only survivor among these patients had radiation preceding amputation 3 months after onset. On the other hand, 4 of these 82 cases were judged to be of relatively low malignancy. All 4 had early amputation, and all 4 died promptly with metastases. It ...

Since 1939 the author has recommended avoidance of prompt amputation early in the disease. Of the many cases in which the advice was not followed (usually because someone concerned *knew* that immediate ...

* Brookline, Mass.

in which the advice was acted upon are included in the case reports that follow, and the results justify emphatically the attempt to find some treatment more satisfactory than prompt amputation early in osteogenic sarcoma.

## THE NEWER METHODS OF TREATMENT

Chemical treatments of many kinds and also some immunity reactions are under study but have not yet shown the capacity to yield satisfactory results. While they do not yet justify selection as the method of treatment, they do merit further investigation, particularly in some cases which already have metastases or have refused surgery or have lesions not accessible for radical surgery.

Three cases of unusual interest in connection with possible immunity reactions are mentioned below; two of them belong in the series about to be reported.

A Registry case records a boy having amputation of the leg followed by metastases in the lungs. He coughed up tissue that was identified as osteogenic sarcoma. This case was believed to be hopeless at the time, and the boy had no further treatment. Five years after amputation he was alive and well with no evidence of pulmonary metastasis. Apparently the metastases had resolved spontaneously.

Case 1.[3] A boy, aged 18, had sclerosing osteogenic sarcoma of the proximal portion of the tibia At biopsy, the tumor was excised and the defect was filled with bone chips. Radiation of the area then was begun and continued until amputation in the seventh month of the disease. The boy soon developed pulmonary metastases, but these reached a static condition with no further development of indentifiable metastases and with the patient in reasonably good health 3 years after amputation

Case 2. A female, aged 21, had onset of disease on September 20, 1945, with biopsy on October 19, 1945, showing rapidly growing osteogenic sarcoma Radiation 3,500 r was given from November 9, 1945, to January 10, 1946, and amputation was done at the end of the fourth month of the disease. In 1947 there was metastasis in the right chest wall. It was radiated and excised in December. It recurred, with spread to an axillary lymph node in 1948. After radiation, a block resection including the recurrence, the ribs and the axillary nodes was performed. The patient was alive in 1958, 13 years after onset and 10 years after the last surgery, but intense itching of the skin had begun in 1947 and multiple skin nodules had developed. No surgeon was ever persuaded to remove one of these for diagnosis. They still were persisting in 1958, although there had been no recurrence of the treated lesions and no pulmonary metastasis. If the nodules were metastatic, had there been an immunity reaction that limited them?

These 2 cases suggest that, eventually, immunity studies in osteogenic sarcoma may be rewarding, but the improved results from avoiding prompt amputation early in the disease are believed to be due not to an immunity reaction but to restriction of surgery to a radiation-induced quiet phase of the disease, as suggested by Ferguson[3] and as illustrated in some of the cases that follow.

### Roentgen Radiation Alone

Roentgen therapy is a useful agent in the treatment of osteogenic sarcoma Currently, its most obvious field of usefulness is as a preliminary to surgery that will follow promptly after completion of the radiation. Apparently, rarely has radiation alone eliminated the tumor, but it is the best available treatment when surgery is not feasible or has been refused.

Case 3. A female, aged 48, had onset of disease at the upper humerus on October 1, 1943 Biopsy on December 18, 1943, showed osteogenic sarcoma. Amputation was refused. Radiation 6,600 r from December 20, 1943, to February 25, 1946, kept the lesion under satisfactory control, but after that it could not be controlled. Ulceration developed, and the smell of the wound finally resulted in consent to amputation, which was done on July 19, 1946, in the third year of disease. Atypical pneumonia was contracted in the summer of 1947, pulmonary metastases were evident in September, and the patient died on December 12, 1947, in the fifth year of disease and 17 months after am-

putation. No effective immunity developed in this case.

Case 4. Green[7] reports a female child with osteogenic sarcoma of the upper tibia who was treated by radiation 9,000 r with rotation technic at 2 million volts. Eventually, an amputation was performed elsewhere because of radiation effects in the leg, and no tumor was found in the amputated specimen. The child was reported to be well 6 years after onset. It is possible that, eventually, such supervoltage technic may prove to be more effective than the more commonly used 200 to 250 kilovolts.

Green[7] reported 5 other cases treated by radiation alone. Of those, 3 are dead, 1 is alive with metastases, and 1 is alive with no known metastasis. Obviously, at present radiation alone is not a sufficiently satisfactory treatment, although it deserves further trial, and its use is practically compulsory when surgery is not acceptable or not applicable.

### Surgery Without Radiation

Avoidance of prompt amputation early in osteogenic sarcoma by performing a more limited operation such as excision initially or by simple delay of amputation[4] has not been as satisfactory when no radiation was used as when the operation was performed at the completion of preliminary radiation. Nevertheless, the following cases are reported so that there will be no omissions in the list of cases in which the author knew in advance that prompt amputation was to be avoided.

In 2 cases of osteogenic sarcoma near the knee, excision was performed with amputation 1 month later. One died with metastasis in the first year; the other apparently was well 2 years afterward, but the further history is unknown. The idea that radiation before operation might be helpful was rejected in these cases.

A girl with osteogenic sarcoma of the lower femur was an interesting case of delayed amputation but not one in which the author was concerned[1] At a large tumor clinic the parents were told that immediate amputation was the only procedure that could save the girl's life. This being refused, she spent 5 years at home with no treatment and then returned to the clinic with a massive tumor in the thigh but no metastases. As this was before the days of hemipelvectomy, the parents were told that the tumor was too extensive to permit surgery. Amputation was sought and obtained elsewhere, and the girl apparently was well 5 years later, 10 years after onset of disease.

### Lobectomy

Lobectomy has been used with success in the treatment of a single pulmonary metastasis, but it will be many years before the probability of success for such treatment is known. The question as to whether or not radiation should precede lobectomy must be considered. Radiation is not recommended by the author for this purpose or for prevention or elimination of pulmonary metastases; he believes that radiation sufficient to eliminate cells of osteogenic sarcoma will result in radiation changes that will be fatal, and a lesser degree of radiation will produce radiation changes that may facilitate development of metastases from surviving cells.

### Limited Surgery After Radiation

Case 5. A male, aged 36, had onset of osteogenic sarcoma at the lateral portion of the clavicle in January, 1954. After biopsy in the second month, radiation 3,600 r was spread over 8 weeks to May 1, 1954. The outer two fifths of the clavicle was resected promptly afterward. At the time of writing, the patient is well with no evidence of disease in the fifth year after surgery and 5 years after onset.

Case 6. A male, aged 34, had onset of osteogenic sarcoma on April 15, 1944, at the lower left femur with amputation on July 20, 1944, in the fourth month of disease. In the fourth year after amputation he developed metastasis at the medial condylar area of the right humerus after an incident in which strain was suffered at support. Radiation 3,200 r was given before excision and implant of bone graft in March, 1948. After local recurrence of tumor the arm was amputated in October, 1949. The patient remained well with no evidence of disease in 1958, 9 years after the second amputation, 14 years after the first amputation, and in his fifteenth year after the onset of the disease. The

sels in an inert state not sufficiently well nourished to multiply to form a metastasis, and that when an incident occurs which aids them to escape from the vessels into an area of clotted blood (or stagnant serum), their nutritional state is improved, they multiply and form a metastasis. Such an incident was the strain to the right shoulder mentioned in this case.

Case 2, described earlier, ...

metastasis in the chest ...

lowed promptly by surgery is preferable to

by prompt amputation early in the disease ...

been designated as "selective amputations."

level of elevated serum phosphatase toward the normal level. Ferguson, in 1950, ...

those are included in Green's report and are not described individually here in order to avoid duplication. In 1950, 3 of those 4 cases appeared to be well (1 of them in the fourth year after amputation), and the fourth was alive but with metastasis suspected.

Case 7. A female, aged 15, had onset of

1946, and death occurred on June 20, 1946, in the second year after amputation; the third year of disease.

Case 8. A female, aged 30, had onset of

23, 1945, radiation 7,100 r was administered, but amputation was not permitted till the patient's husband returned from war service. It

patient took numerous airplane trips before and

*Obviously these 3 cases do not represent* the optimum handling of selective amputation. Two of them died within 2 years, and 1 was apparently well 5 years after amputation. Three cases more nearly optimum in their handling follow.

*Case 10.* A male, aged 36, had onset of osteogenic sarcoma at the lower tibia on December 7,

1940, and was seen on December 10, 1940. Radiation 4,400 r was given from January 27

developed pulmonary metastasis in the sixth year and died in that sixth year

been permitted

1946, and January 23, 1947, with amputation on February 3, 1947, in the sixth month of disease. There was no evidence of disease in 1958, 12 years after onset and 11 years after amputation.

These 3 cases, seen fairly early and, after radiation, amputated fairly early, show 1 death in the first year and 2 patients alive and apparently well 5 years after amputation. Of these 2, 1 developed metastasis and died in the sixth year, and the other is alive and apparently well

Cases 1 and 2, previously described, and Cases 5 to 12 are examples of selective surgery, although Case 6 had radiation before the surgery only when the metastasis was treated, and in Cases 7 and 8 the amputation was delayed longer than intended

desired. The results relative to the 10 cases

2 alive but not well 3 and 13 years afterward respectively.

Of 7 patients after 5 years:

4 well and apparently free of disease 5, 6, 12, and 15 years afterward, respectively.

1 well 5 years afterward but dead 1 year later.

3 dead

some delay and possibly the errors. The author personally radiated Cases 2, 5, 6, 10 and 12 (excluding Case 3, which was not a selective amputation because the operation was performed very late and in an active stage of the disease). The results in these 5 cases were:

1 alive but not well after 13 years

48-hour intervals, decreasing to 2 treatments weekly with 48-hour intervals when the lesion appeared to be becoming quiet, and continuing the treatment through 8 weeks with amputation in the following week. Had there been recurrence of activity of the tumor, the treatment would have been begun

DR. GREEN'S CASES

Green's report includes 11 cases of selective amputations. None was adult. Radiation was more intense and less prolonged than in the cases radiated by the author. Possibly this is why the results are better, especially in younger cases; seven patients were still in their third year. The remaining 10 cases are summarized here in order to make a consolidated report on the results of selective amputation. Four of the 10 cases had

previously been mentioned by Ferguson,[6] and 6 were more recent. The results reported were:

Of 7 patients after 5 years or longer, 4 apparently well, 3 dead.

Of 3 patients after 2 but not 5 years, 2 apparently well, 1 dead.

CONSOLIDATED REPORT

Combining the above 10 cases from Green's list with the 10 cases of selective surgery in the author's list, the results as presently known are as follows:

4 cases more than 2 but less than 5 years after onset:

1 alive with metastasis.
2 well with no evidence of disease.
1 dead.

16 cases more than 5 years after onset:

1 alive but not well after 13 years.
8 well and apparently free of disease.
7 dead including 1 who apparently had continued well for more than 5 years.

The results of selective surgery appear to be much better than those of prompt amputation early in osteogenic sarcoma. The author believes that the principle of selective amputation is also applicable to the treatment of chondrosarcoma, but nothing that has been said here is intended to apply or believed to apply to the treatment of any other variety of tumor, including Ewing's sarcoma, fibrosarcoma and solitary myeloma.

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## Tendentias Moderne in le Tractamento de Sarcoma Osteogene

*Summario in Interlingua*

Recente evenimentos indica que on ha generalmente recognoscite que le resultatos del prompte amputation in un stadio precoce de sarcoma osteogene es si magre que le essayo de altere formas de tractamento es indicate. In un revista publicate per Ferguson in 1940, le sequente statistica, que es typic, esseva presentate: Un gruppo de dece-tres patientes operate durante le prime mense post le declaration del symptomas includeva nulle superviventes; un gruppo de vinti-cinque patientes operate durante le secunde mense includeva un supervivente; un gruppo de vinti-tres casos operate durante le tertie mense includeva duo superviventes.

Plure methodos de tractamento ha essite essayate in loco del prompte amputation in stadios precoce de sarcoma osteogene. Reactiones immunologic e methodos chimotherapeutic ha non ancora demonstrate un utilitate sufficiente, sed illos merita investigaciones additional. Radios X como sol forma de tractamento ha sporadicamente producite bon resultatos sed non satis frequentemente pro establir irradiation como therapia de election, excepte in casos in que interventiones chirurgic es impracticabile o refusate.

Le uso de roentgeno-irradiation durante plure septimanas sequite promptemente per

le intervention chirurgic (amputation in tanto que possibile) ha producite resultatos que justifica le uso de iste forma de tractamento in loco del prompte amputation como therapia initial in le prime sex menses post le declaration de sarcoma osteogene. Isto es indicate in un recente reporto per William Green ab le Hospital Infantil de Boston e etiam in un reporto publicate per Ferguson in 1950. Iste ultime reporto es augmentate e ponite al die in le presente publication.

Le radiation administrate per Ferguson amontava a inter 3200 e 4000 r de alte voltage distribuite super un periodo de inter sex e octo septimanas, sequite per le intervention chirurgic intra dece dies post le ultime tractamento. In le casos de Green, le radiation esseva un pauco plus accelerate e plus intense. Illo amontava a inter 4000 e 5000 r in inter quatro e sex septimanas, e in certe casos radiation a supervoltage esseva usate.

Le resultatos in le duo series esseva similissime. Proque quatro casos es includite tanto in le un como etiam in le altere serie e etiam proque le numero total del casos es basse, le duo series es consolidate pro exprimer le sequente resultatos:

Inter dece-sex casos, octo se trovava ben cinque annos post le operation; un non se trovava ben; e le remanente septe patientes esseva morte. Un del casos de Ferguson, que se trovava ben post cinque annos, disveloppava metastases durante le sexte e moriva. Con iste exception, omne le casos que se trovava ben duo annos post le operation se trova ben ancora a periodos de inter cinque e dece-cinque annos post le operation. Le caso que non se trovava ben cinque annos post le chirurgia (etiam in le serie de Ferguson) vive a iste tempore, i.e. dece-tres annos post le tractamento initial. Durante plus que dece-un annos iste patiente ha habite un abundantia de nodulos cutanee con intense prurito.

Le casos con minus que cinque sed plus que duo annos passate depost le operation include duo que se trova ben, un que non se trova ben, e un que es morte.

Le resultatos justifica essayos additional del tractamento de precoce sarcoma osteogene per roentgeno-irradiation sequite promptemente per le intervention chirurgic como methodo preferite al immediate amputation initial.

# Cerebral Palsy: The Upper Extremity*

FRANK H. STELLING, M.D. AND LESLIE C. MEYER, M.D.

The habilitation of the upper extremity in the cerebral palsy patient presents a difficult and often frustrating problem. Great [illegible]

[illegible] tremor, remembering that mixed types can, and do, occur.

Treatment is made more difficult because the majority of these patients have never previously experienced normal functional patterns. Rehabilitation of the upper extremity in the cerebral palsy patient who previously had a normal extremity is much easier than the habilitation of the child afflicted from birth Frequently, mental retardation is present, and, when combined with possible hearing and visual defects, the problem may be insurmountable.

The treatment of the early-age group, usually considered the preschool group, is

[illegible] ataxias and the athetoids, contractures are [illegible]

[illegible] der of primary importance. The person doing the exercises is taught to carry each joint [illegible] the d[illegible] attitude. Sometimes it is difficult to get parents to understand and accept this responsibility, so that periodic supervision by the doctor and a trained physical therapist is necessary. It is important to encourage the parents and to see that the exercises are done properly. The exercises must be simple, and their recommended frequency should be realistic so that they can be fitted into the parents' schedule.

As a child grows it may be noticed that certain deformities are developing in spite of the exercises. Then, simple splinting in the form of casts or Celastex molds can be used. These are advised in the young child because they fit snugly and do not rotate as do conventional type braces.

As an awareness of the hand develops, simple active exercises may be started. Oc-

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The authors are indebted to Dr George L Ford, Fellow, Duke University, Orthopaedics Training Program, for his assistance with the References.

cupational therapy then can be used to advantage, and the parents are instructed by an occupational therapist under the direction of the physician concerning the proper use of these facilities. Parental assistance is used to full advantage, but supervision by trained personnel at periodic intervals is important. A careful liaison must be maintained among physician, occupational therapist and physical therapist to keep the patients properly instructed and to develop new methods of instruction as the needs arise.

Restriction of the normal or dominant hand should never be permitted to emphasize the use of the involved extremity because of the possibility of producing serious emotional disturbances. In training the athetoid or the spastic quadriplegic patient, Deaver recommends the elimination of all but two movements, thereby making it possible to teach the child many self-care activities; for example, by tying the arm to the body and placing a cock-up splint under the wrist, the movements of the upper extremity are limited to flexion and extension of the elbow. [illegible] tered, [illegible] contro[illegible] be pos[illegible] wrist, th[illegible] planned [illegible]

cerned [illegible] the sup[illegible] muscles [illegible] much [illegible] though [illegible] occur [illegible] strengt[illegible] tary m[illegible] it necessary for braces to be stronger and more rigid than those used in the flaccid paralysis alone. Also braces should be heavy enough to help steady the extremity, particularly in the athetoid. Generally speaking, dynamic splints using springs and rubber bands are difficult to use and are not suited to the treatment of the cerebral palsied hand, especially of the athetoid. The splints and the braces should be simple, sturdy and easy to apply, and they should control only one or, at the most, two joint motions. An example of this would be the simple bar splint with thumb-extension control. This splint is sturdy and easy to apply, and it controls the gross motion of two joints—the wrist and the thumb. The use of braces or rigid splints such as plaster bivalves or Celastex splints should always be used on the hand before any attempt is made for surgical reconstruction. It is much easier to discard a poorly selected brace than to attempt the correction of a premature and poorly planned surgical procedure. In a spastic hand, distribution of overactive muscles varies with each hand. A typical example is a spastic hand in which the fingers can be used freely when the wrist is held firmly [illegible]

[illegible] by stretching them. The early use of this brace

allows for the gradual correction of the deformity and prevents the development of permanent contractures. It is also important to remember that, in spastics particularly, bracing must be used at night. Any amount of training during the day will be nullified by allowing this extremity to assume a flexed or a deformed position during sleep.

## SURGERY

Usually, the typical spastic upper extremity assumes a position in which the shoulder joint is rotated internally and adducted. The elbow joint is in a position of flexion with the forearm in pronation. The wrist and the fingers are in flexion, and the thumb may be in a position of flexion and adduction (thumb-in-palm position). Some modifications of these positions can occur. In the athetoid, these positions may be present for a time, but they change quickly, and no constant pattern can be formulated. This is well demonstrated by the athetotic phenomena of *spasmus mobilus.* Surgery, if it is justifiable, must tend to correct these deformities and improve the function of the hand or place it in a better position so that it *can* function.

"The majority of hand deformities resulting from cerebral palsy do not warrant surgery."[15] Of 800 patients up to the age of 18 years who were examined at the North Carolina Cerebral Palsy Hospital during a period of 3 years, 300 were found to be eligible for general treatment after consideration of their mental and physical condition. However, only 12 patients (4%) of the selected 300 were considered for hand surgery by Goldner.[11]

McCarroll[17] found that, in general, surgical treatment of deformities of the upper extremity was quite unsatisfactory, and frequently the improvement obtained is cosmetic rather than functional. In 1,459 patients with lesions of the pyramidal tract, only 125 operations were performed on 546 suitable cases, and of these only 16 yielded a result that could be classified as good.

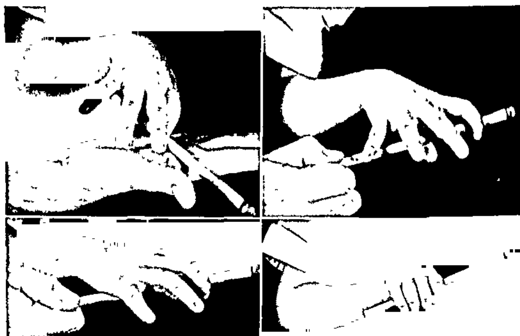


FIG 1. Typical reach-grasp mechanism in spastic type of cerebral palsy showing supplementary reinforcement of finger extension by reaching with the wrist flexed and closing with the wrist extended If the patient can do this well, it is best not to perform surgery. (Stelling, F H., & Ferguson, Albert, Jr : Orthopedic Surgery in Infancy and Childhood, Baltimore, Williams & Wilkins)



FIG. 2. Inability to extend wrist. May be improved by wrist arthrodesis, but passive splinting of the wrist should be used first.

Surgery of the upper extremity in all types of cerebral palsy must be contemplated carefully. Several prerequisites are as follows:

1. Prolonged observation by a trained surgeon is necessary. No surgical procedure should be attempted until the extremity has been subjected to bracing, splinting and a careful evaluation of the patient's potentialities.

2. Generally speaking, surgery of the upper extremity before the age of 8 years is not advisable.

3. Very few, if any, procedures are adaptable to athetoids, rigidities or tremors. Best surgical results are obtained in the spastic. Limited surgical procedures applicable to athetoids are the stabilizing ones.

4. Muscle evaluation, although difficult, must be done carefully and repeated.

5. Sufficient intelligence should be present to ensure co-operation.

6. Surgery may be considered on the useless hand for cosmetic reasons alone.

Intelligence is an important prerequisite for surgery, particularly in tendon-transplantation procedures. A definite level cannot be set dogmatically, but usually there must be a desire for improvement of hand function, together with the ability to co-operate in any reconstructive procedures. Goldner[11] generally chose patients for surgery with an intelligence quotient of about 70, although this particular level was not adhered to if certain specific functions could be accomplished by doing the surgery. Surgery was reserved for those patients with enough intelligence to allow them to follow through with re-education and with enough initiative



FIG 3. Positive plaster mold made with the wrist extended. Plaster splint made over the positive mold so that the splint fits snugly to avoid slipping.

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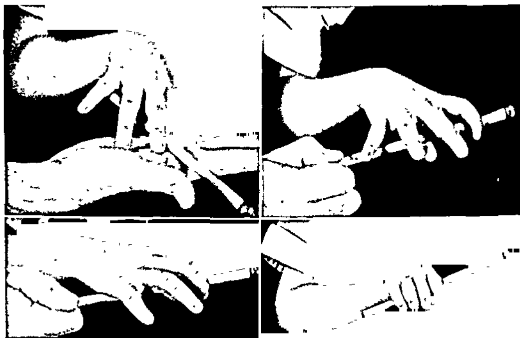


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FIG 3. Positive plaster mold made with the wrist extended. Plaster splint made over the positive mold so that the splint fits snugly to avoid slipping.



FIG. 4. Active supinatory function after transplant of flexor carpi ulnaris to radial styloid (Steindler). This removes ulnar flexion deformity and adds supination power.

to take advantage of the anatomic improvement.

As previously stated, most procedures are carried out on a spastic extremity, and, unless otherwise indicated, the following discussion applies to those cases classified as spastic or the mixed type. Surgical procedures available consist of tendon transplantation, neurectomies, tenotomies and tendon lengthenings, fascial releases and arthrodeses.

It is emphasized that we cannot expect the type of success from tendon transplantations that we obtain in the paralytic deformities. Phelps[21] states that good long-term results cannot be obtained from muscle transplantations in either the spastic or the athetoid patient because the injury in cerebral palsy does not involve individual muscles, as in poliomyelitis or other lower motor neuron lesions, but affects total joint function or position, which is mediated through the upper motor neurons. In the cerebral palsy extremity we are dealing with paralyzed and weakened muscles, but frequently the antagonistic muscles are spastic and unpredictable in their action once they are transplanted to their new position. It is important to study the hand carefully and identify the characteristics of each muscle prior to any type of transplantation

## NEURECTOMIES

Neurectomies about the hand and the forearm have been found to be disappointing, particularly when used alone. A procedure employed frequently was the pronator neurectomy. A review of these cases over a period of 10 years at the Shriners' Hospital, Greenville, S.C., has proven them to be of little value. Selective neurectomies of the ulnar and the median nerves are disappointing because it is difficult to section the correct number of nerves to control the spasticity and also because the finger and wrist extensor power fails to return.

## STABILIZING PROCEDURES

Stabilizing procedures are much more successful in improving function, particularly about the wrist and the thumb. These procedures may be combined with tendon transplantation. The order of these two procedures is considered to be important, but no dogmatic statements can be made. We have found preliminary bracing to test the possible success of a procedure, then stabilization and, finally, transplantation of tendons to improve motor function to be most successful. Goldner[11] advocates tendon surgery as the initial procedure following this with the necessary joint arthrodeses. Cooper[6] believes that there is a latent capacity of the spastic hand that generally is underestimated. He states that after the wrist is fused successfully, ordinarily gradual improvement in active extension of the fingers develops without assistance from splints, stretching or surgery.



FIG. 5 (*Top, left*) Result sometimes attained after transplanting wrist flexors to finger extensors. Loss of flexor stability of wrist. May be avoided by leaving at least 1 wrist flexor. (*Top, right*) Showing patient's ability to extend fingers if flexor stability is added. Patient should have wrist arthrodesis in this position. (*Bottom, left and right*) Reach and grasp following good wrist arthrodesis. Wrist in ulnar deviation enhancing thumb abduction and extension. (Stelling, F. H., & Ferguson, Albert, Jr.: Orthopaedic Surgery in Infancy and Childhood, Baltimore, Williams & Wilkins)

## Fasciotomy and Myotomy

Contractures of fascial structures and muscles do occur, particularly in the older-age groups. These should always be corrected by adequate sectionings. Tenotomies and tendon-lengthening procedures will be of value in a very small number of cases. It may be stated generally that tenotomies and tendon-lengthening procedures have proven to be more successful than neurectomies in controlling an overactive spastic muscle. One reason for this is because neurectomies depend, to a large degree, on the surgeon's judgment. It is most difficult to evaluate the degree of spasticity of a muscle and cut enough of the nerve supply to improve its function. It is agreed universally that too little should be done rather than too much. Stabilizing procedures are more successful than tendon transplantations because muscles transplanted are spastic and, therefore, unpredictable in their new location. Re-education of the spastic muscle often is difficult, if not impossible.

Now we can turn to more specific observations concerning surgical procedures of the upper extremity. Contractures about the shoulder and flexion deformities of the elbow usually do not warrant any type of correction. Occasionally the Sever operation may be employed to correct a severe adduction internal rotation contracture. The choice of this operation would depend entirely on improving the usefulness of an already satisfactorily functioning hand. It could be of aid in improving axillary hygiene.

The elbow is usually in some degree of flexion, and this, in itself, is a position of choice. Tenotomies or lengthening operations of the biceps brachii are rarely, if ever, necessary.

## Surgery for Pronation Deformities

Most of the surgery about the forearm is designed to correct the severe pronation deformities. An operation often employed to assist in correcting this deformity and to improve supination is the pronator neurectomy. This operation is unsatisfactory if used alone; it should be combined with other procedures. The pronator teres is a powerful muscle, usually very spastic, and can be transferred into the extensor carpi radialis longus, thus supplementing wrist extension. When this is done, it is advisable to consider transferring the flexor carpi ulnaris into the

extensor carpi ulnaris to prevent radial deviation of the wrist. The technics of Steindler[26] and Green[12] in the transplantation of the flexor carpi ulnaris have been useful in removing the flexor carpi ulnaris as a producer of the flexion-ulnar deviation deformities of the wrist and using it to help correct spastic pronation deformity. The two procedures differ in that Steindler transplants the flexor carpi ulnaris directly into the distal end of the radius; Green transplants the flexor carpi ulnaris into the extensor carpi radialis longus, thereby utilizing the tendon transplant to obtain some wrist extension as well as to correct the pronation deformity. With either of these two preceding operations it is advisable to remove the effect of a spastic or contracted pronator teres by its transplantation or denervation and tenotomy. The pronator quadratus should not be ignored as a deforming factor and should be sectioned if it is spastic or contracted. Frequently it is found that all procedures designed to improve the severe pronation deformities fall far short of the desired improvement of function. Often correction of the deformity is the only end-result.

### Arthrodesis of the Wrist

Perhaps the most valuable operation of the entire upper extremity is arthrodesis of the wrist. This procedure corrects a disabling deformity, and the improvement of position allows for better function of the finger extensors. The appearance of the hand is improved and is important in its psychological effect on the patient, particularly the hemiplegic. The success of wrist fusion is largely dependent on the potential muscle power in the wrist and the finger extensors. Occasionally, transplantation of the wrist flexors may be necessary to improve the function of the finger extensors; however, these transplants will improve the extension of only the metacarpophalangeal joint and not the interphalangeal joints.

Successful extension of the interphalangeal joints depends on normally innervated intrinsics, a condition seldom found in the spastic hand. Careful observation of the flexed wrist will reveal that some patients have fairly good function of the hand in the flexed position. This is true because the wrist flexion allows for adequate extension of the fingers. It is inadvisable to attempt to improve this type of hand by wrist arthrodesis and/or transplantation. It must be reiterated that no surgical procedure should be carried out until the surgeon is thoroughly familiar with the patient's capabilities

Technics of wrist fusion vary, and many are successful. Some basic principles that are beneficial to the spastic hand will be considered. Sometimes it is advisable to remove a row of carpals to give more length to the contracted flexor tendons. Total carpectomy and fusion of the radius to the metacarpals may be indicated in severe cases of flexion deformity and contracture. In those cases with marked flexor spasticity it is advisable to extend the fusion into the metacarpals, obliterating completely any wrist-joint motion. In some hands a few degrees of motion may be necessary to permit a tenodesing effect and to allow for an automatic grasp and release. In these hands it is advisable not to fuse the carpometacarpal joint.

Generally, wrist fusions are advisable in a position of 20° or 25° of extension. Eggers[9] suggests a 35° hyperextension, stating that some of the hyperextension is lost early in the spastic hand and that additional flexion at the wrist occurs during subsequent years.

Goldner[11] has found that, for certain hands, the neutral position of 180° has been satisfactory, while others have functioned best in 5° or 10° of flexion. He believes that in the hyperextended position there is increased tension on the finger and the thumb flexors, thereby increasing the finger-in-palm and thumb-in-palm deformity. Considerable ulnar deviation of the fused wrist is advocated in an effort to help correct the "thumb-in-palm" defomity. Placing the wrist in

ulnar deviation throws stress on the thumb extensors and abductors to help pull the thumb out of the palm. Resection of the distal end of the ulna may be helpful in these wrists in securing more ulnar deviation.

### Thumb-in-Palm Deformity

The "thumb-in-palm" deformity is the second most disabling problem in the spastic hand. Its position prevents grasp, and, because of the spastic musculature, there occur contractures that prevent any useful motion of the thumb. Frequently, if the deformity is of a mild degree, simple ulnar deviation of the fused wrist, as previously described, will place it in a satisfactorily functioning position.

Tendon transplantation used alone about the spastic thumb gives disappointing results. Usually, some type of stabilization of the metacarpophalangeal or the carpometacarpal joints is necessary. Frequently, combining the two procedures will improve gross function, provided that the transplanted tendons can be re-educated. Stabilizing procedures about the thumb vary in type and extent. Bone-grafting between the first and the second metacarpals provides good stability and optimum position, but there is a tendency for the deformity of the thumb to recur with growth. Hyperextension, hyperflexion and instability of the metacarpophalangeal joint occur frequently, making it necessary to fuse the joint secondarily. A satisfactory stabilization can be obtained more easily by arthrodesing separately the metacarpophalangeal and the carpometacarpal joints of the thumb.

Steindler[26] is of opinion that tendon transplantation about the thumb provides no lasting effect and that in time the thumb assumes its original position. He advocates stabilizing the carpometacarpal and the metacarpophalangeal joints in a position of opposition to form a "firm post" against which the fingers of the hand are moved. He reports 50 per cent good and 43 per cent poor results from the use of this operation.

Goldner[11] has advocated transplantation of active wrist or finger flexors to the extensor pollicis longus. He has also recommended rerouting the extensor pollicis longus, thereby decreasing the chance of the thumb's rolling into external rotation and adduction. If the metacarpophalangeal joint of the thumb is unstable, he favors fusion at the same time that the transfer to the extensor pollicis longus is done. In any procedure used to improve the "thumb-in-palm" deformity, it is advisable to eliminate spasticity and contracture of the thumb web by sectioning tight thumb web fascia, stripping the first dorsal interosseous from the first metacarpal and sectioning the adductor pollicis.

Finger function can be improved occasionally by the transplantation of an active motor, such as a wrist flexor, into the extensor digitorum communis. Other procedures advocated frequently for paralytic deformities about the hand have been successful in only a very limited number of cases. They should only be attempted by the experienced hand surgeon after prolonged observation, careful planning and reasonable assurance that the procedure has a fair chance of improving the function.

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## Paralyse Cerebral; Le Extremitate Superior

*Summario in Interlingua*


Le rehabilitation del extremitate superior in paralyse cerebral, e plus ancora le habilitation del nunquam usate extremitate superior in tal casos, es un problema difficile que depende in grande parte del appropriate selection e classification del patientes.

A basse etates, le tractamento de omne classes de patientes visa a render le infante conscie del existentia del mano, comenciante per extension passive e continuante per exercitios active e therapia occupational. Un simple apparato es possibilemente de adjuta in prevenir le disveloppamento de deformitates. Illo etiam servi a eliminar certe movimentos in les trainamento del infante. Restriction del mano normal o dominante non debe esser tolerate a ulle tempore o pro ulle rationes.

Le apparatos usate debe esser plus pesante que illos usate in casos de paralyse flaccide. Lor function debe esser primarimente supportative. Apparatos dynamic es generalmente pauco satisfactori. Le uso de apparatos orthopedic es etiam de valor como adjuta in le evalutation pre-chirurgic del expectate efficacia del intervention chirurgic mesme.

Interventiones chirurgic es de valor in solmente pauc e cautemente seligite casos. A generalmente parlar, illos non debe esser

interprendite ante le etate de octo annos, e mesmo alora rarmente in casos de athetosis, rigiditate, o tremor.

Le melior resultatos chirurgic es obtenite in casos spastic. Le intelligentia del patiente es un factor importante in le determination del successo chirurgic.

Fusion carpal es, per multo, le manovra chirurgic le plus satisfacente. Frequentemente illo pote esser combinate con allongation o transferimento de tendines. Le stabilisation del pollice in un position functional—como "palo de supporto" con omne le articulationes stabilisate o con un combination de fusion e transferimento de tendine—es etiam un manovra satisfactori.

Neurectomias per se es disappunctate, sed in combination con altere mesuras illos es frequentemente utile.

Liberation fascial e tenotomias es de valor in certe casos. Le interventiones chirurgic in le region del cubito e del spatula es extrememente inextense. Illos es justificate solmente in le forma de manovras de liberation.

In le antebracio le majoritate del operationes visa a corriger le deformitates pronational. In tal operationes il se tracta usualmente de neurectomia o de transferimento del pronator terete. Transplantation del flexor carpal ulnari, con le objectivo de facer lo ager como supinator e extensor, es etiam de valor.

Es discutite detalios technic relative al fusion carpal, al fusion del pollice, e al associate transferimentos de tendine.

# 8

# Anteversion of the Femoral Neck

T. GORDON REYNOLDS, M.D., AND FRED E. HERZER, M.D.*

## TERMINOLOGY

Anteversion of the femoral neck is a forward twisting of the femoral neck, actually *external rotation of the upper end* of the femur in relation to the lower end, or an *internal rotation of the lower end* in relation to the upper end. The term *femoral torsion* is also used when referring to this relationship between the lower and the upper ends of the femur.

The angle of anteversion of the femoral neck could be defined arbitrarily as the angle between two planes (Fig. 2, I) that intersect in the longitudinal axis of the femoral shaft, one passing through the neck and the center of the head and the other parallel to the transverse axis of the condyles (and passing posteriorly to the center of the head of the femur). If this transverse plane passes anteriorly to the center of the femoral head, then retroversion is present.

In adults, normal anteversion is about 8° to 15°; in children 1 year old, it may be as much as 50° (Somerville;[17] Budin & Chandler[2]). Shands and Steele[16] found that infants up to 1 year had an average of 39°; at 2 years it was 31°, with gradual decrease to 24° at 10 years; and at 16 years it was 16°. With weight-bearing, anteversion begins to decrease.

Both the cervicofemoral angle and the amount of anteversion may become altered by abnormal stresses from muscles, ligaments or weight-bearing, as in congenital dislocation of the hip, poliomyelitis, cerebral palsy and improper habit posture of sitting or sleeping.

While it is impossible, even on a dry femur, to measure the amount of anteversion precisely, because of the difficulty in drawing straight lines and angles that approximate the complicated curves found in nature, a fairly accurate knowledge of the amount of anteversion is of great importance in the treatment of children with congenital or paralytic dislocation of the hip. It is also of importance to know the amount of anteversion in such conditions as coxa vara, coxa plana, clubfeet and other developmental or metabolic abnormalities of the lower extremities. At times, gait and posture problems are also associated with abnormal anteversion. If a rotational osteotomy is to be used to correct excessive anteversion, it is helpful to know as accurately as possible the amount of anteversion present.[6,10,12,14,18]

Routine anteroposterior and frog-leg roentgenograms of the hip (Figs. 3 & 4) can show the amount of anteversion of the femoral neck, if taken with proper positioning of the extremities. By its very simplicity and ease of application, this method of determining anteversion will lead to a better understanding of the relationship of anteversion not only to congenital dislocation of the hip but also to the many other conditions affecting gait in children.

In recent years there have been published several papers describing various

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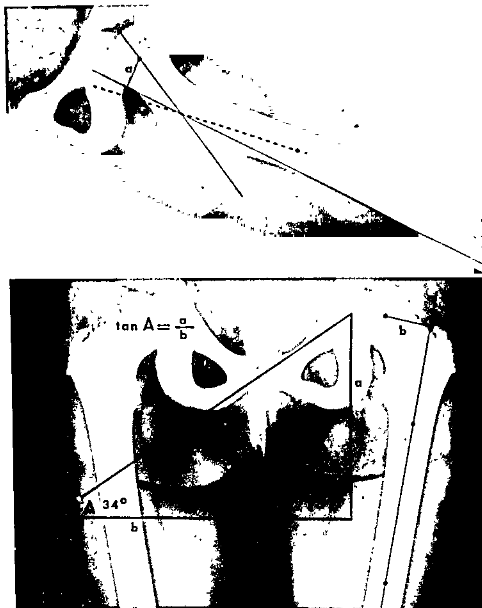



FIG. 1. B. D., 4 years old. Frog-leg lateral and anteroposterior views of upper femur. "a" shows posterior displacement femoral shaft and "b" the lateral displacement in relation to center of head. A right triangle with sides *proportional* to "a" and "b" illustrates amount of anteversion angle "A," opposite side "a."

NOTE: The dotted line (lateral view upper) represents only the axis of the upper end of the shaft, while the solid line is the axis of the entire shaft.

Use the solid line for measuring anteversion. It is more accurate, especially when thighs are not fully abducted.

Using broken line to measure "b" would make angle "A" 40° instead of 34°

methods of determining roentgenographically the amount of anteversion of the femoral neck.[1-5,7,11,13,15,18] The special positioning required in most of these cannot be adapted to routine anteroposterior and frog-leg lateral roentgenograms taken ordinarily of the hips.

## RIGHT-TRIANGLE METHOD OF DETERMINING ANTEVERSION

If someone tells you that he lives 6 blocks east and 6 blocks north of a certain spot (Fig. 2, III), then you know that he lives 45° north of east of it. Similarly (Fig. 2, II), the axis of the femoral shaft can be thought of as being displaced posteriorly and laterally to the head. These distances can be measured on true lateral and anteroposterior roentgenographic views. Figure 2, II is a diagrammatic representation that shows both "a" posterior and "b" lateral displacement of the femoral shaft if projection could be made in line with the axis of the shaft.

The procedure is as follows:

1. Measure on the frog-leg lateral projection (Fig. 1, *top*) the distance "a" from the center of the head to the extended axis of the femoral shaft This shows the posterior displacement of the shaft (Fig. 2, II).
2. Measure on the anteroposterior view

I

Axis of femoral neck

Angle of anteversion A=

Axis of femoral shaft

Transcondylar axis

II

a

b

50°

L C INNES

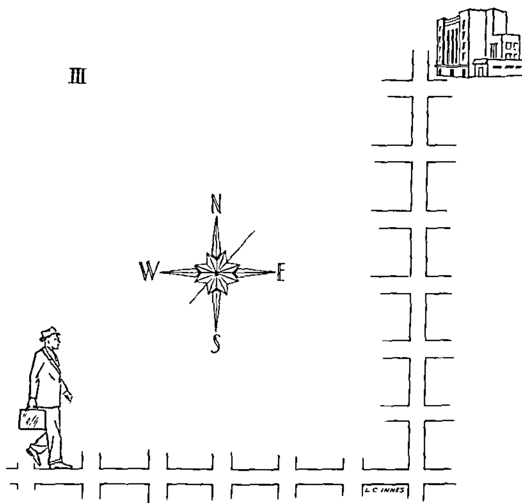



Fig 2 (*Top*) I—Anteversion of femoral neck is the angle between two planes, both through the shaft and one through the neck and the head, the other transcondylar. II — In axial view of femur angle between neck and transcondylar plane is "A," the anteversion "a" posterior and "b" lateral displacement of shaft also shown. (*Bottom*) III — Direction determined by two measurements taken at 90° to each other.

(Fig. 1, *bottom*) distance "b" from the extended axis of the femoral shaft to the center of the head. This gives the lateral displacement of the femoral shaft from the head (Fig. 2, II).

3. Draw a right triangle (Fig. 1, *bottom*) with altitude and base *proportional* to "a" and "b." Measure angle "A" opposite side "a"; this is the angle of anteversion.

*Caution.* Do not measure the wrong angle on the right triangle. Angle "A," the anteversion, is opposite side "a," and side "a" is the posterior displacement shown on the frog-leg lateral. Is the result obtained reasonable? If the femoral shaft is more lateral ("b") than posterior ("a"), anteversion will be less than 45°. If "a" is greater than "b," then anteversion will be more than 45°.

If, instead of drawing the triangle (Fig. 1), you prefer a slide rule, the formula that

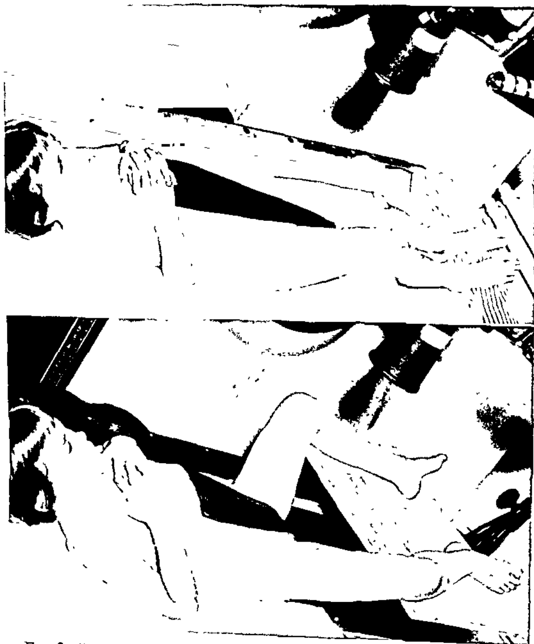


FIG 3. (*Top*) Position for anteroposterior view of hips. Note gonad shield and support under knees to prevent external rotation. (*Bottom*) Position for "frog-leg lateral." Note adjustable platform. Knees flexed 90°. Legs horizontal.

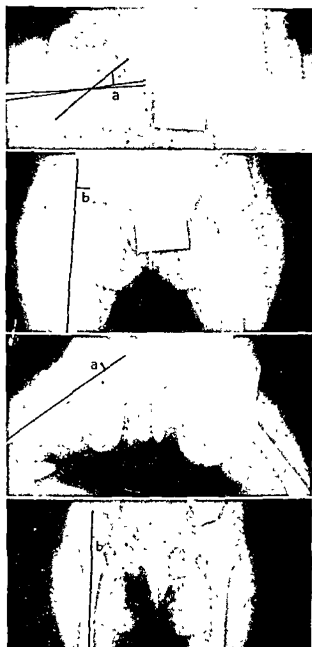



FIG 4. (*Upper two illustrations*) A. B., 10 months, anteversion 45° (*Lower two illustrations*) D R., 4 months, anteversion right hip 60°.

probably was the first that we learned in trigonometry may be used:

$$\tan A = \frac{a}{b}$$

POSITIONING

Proper positioning is necessary for accurate results.

It is advisable to protect the gonads by a shield that does not obscure the hip joints.

The anteroposterior view of the femoral neck as shown in an ordinary flat of the pelvis (Figs. 1, *bottom*, & 3, *top*) with the femur in the *anatomic position* is adequate because it brings the transcondylar axis into a horizontal plane parallel to the table top. Any other position that keeps the transcondylar axis of the femur in the horizontal plane can also be used. Flexion does not affect the value of "b," but rotation would. Actually, accuracy is facilitated by tying the knees together and flexing them about 20° over a support.

The so-called frog-leg lateral of the hips (Figs. 1 & 4, *top*, & 3, *bottom*), to be true lateral, must have two specifications. *The knees are bent to a right angle, and the tibias are held horizontally.* The thighs are abducted as far as is comfortable. The more the abduction the greater the accuracy. It can be shown geometrically that this position—*the knees flexed to 90° with the tibias in the horizontal position*—rotates the femur 90° on its long axis in relation to the axial x-ray so that the transcondylar plane is now perpendicular. Positioning the frog-leg view is facilitated by supporting the legs on an adjustable platform

*Note.* Theoretically, the amount of abduction in either a vertical or a horizontal axis has no effect on the distance "a."

This variation, permissible in positioning, often makes it possible to place the femur properly for a true lateral, even when contractures are present. Calculations are more accurate, especially in small children, if the entire length of the femur is included in the picture.

It is interesting to note that if a frog-leg lateral view is positioned properly with the x-ray tube centered over the upper end of the femur, regardless of the amount or the direction of abduction, the roentgenogram of the knee will depict a right angle.

It should be noted that "true lateral" as sed here means only that the transcondylar xis is in a vertical plane common to the xial x-ray. Error due to the centering of the ube between the femoral heads instead of ver the head is slight and can be disregarded.

Proper positioning is of the utmost importance. If the technician understands horoughly the principle involved, error from positioning can be reduced to a minimum.

### Where Should the Lines Be Drawn?

On both anteroposterior and lateral projections the center of the head is marked with a small dot (Figs. 1 & 4). A pair of dividers can be used to locate the center more accurately.

In children in whom the epiphysis has not ossified, a point midway across the metaphysis will approximate closely the center of the head (Fig. 4).

1. On the lateral view (Figs. 1, *top,* & 4, *top*) a line drawn along the posterior cortex of the middle one third of the femur and extended proximally to the level of the head will pass through the base of the femoral neck and will represent the axis of the femoral shaft. This line would also pass through the mid-line of the lower end of the shaft.

Measure distance "a" from this line to the center of the head.

If greater precision is desired, the "base of the neck" can be located as follows (Fig. 1, *top*):

A Draw a line through the center of the head and through the mid-line of the neck.

B. Place a dot in the mid-line of the upper one third of the femoral shaft at the narrowest portion. Extend a line proximally from this point parallel to the anterior cortex. Its intersection with the line through the center of the neck is the "base of the neck." The axis of the femoral shaft should pass through this point.

2. On the anteroposterior projection (Fig. 1, *bottom*) place two dots in the mid-line, one at the upper and the other at the lower end of the shaft. Extend a line through these proximally to the level of the head. This line represents the axis of the femoral shaft.

Measure distance "b" from this line to the center of the head.

For more precise results use a fine-line wax pencil. Wax-pencil markings can be changed or removed later if desired. We have found that, instead of marking on the film, a strip of cleared x-ray film on which a long straight line has been placed is more accurate. This can be superimposed on the film and moved easily one way or the other until the exact position desired is obtained. Measurements then are taken before the strip is moved. A horizontal view box is more convenient than a vertical one in taking these measurements.

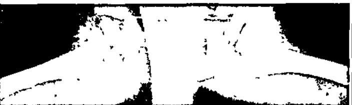
### Degree of Accuracy

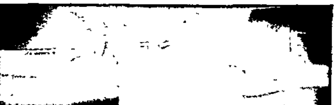
The above method of measuring anteversion of the femoral neck is recommended after a careful study of the skeleton. Actual anteversion was measured by visual inspection of dry femurs and roentgenographic axial views of the same bones. Anteroposterior and 10°, 30° and 90° frog-leg views also were taken. By drawing the lines as described above, the calculated anteversion on these views was within ±3° of the average of measurements on the axial x-ray projection. The 10° abduction view showed the greatest deviation. In clinical use an error less than ±10° is expected.

### Estimation of Anteversion by Inspection Only

Probably the most frequent use of this method is that, *once the principle is understood thoroughly*, the amount of anteversion can be estimated roughly by only a glance at *these special* anteroposterior and frog-leg views.

1. *There is about 45° of anteversion* if both views show the head to be about the





I

II

III

R



IV

V

VI



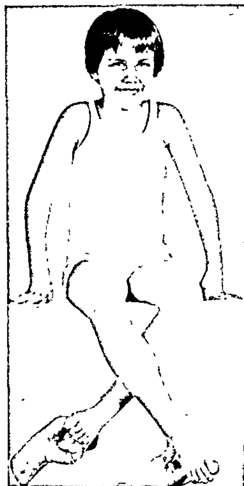


FIG. 6. (*Above*) Maximal internal rotation. (*Right*) Maximal external rotation.

same distance from the axis of the femur (Fig. 5, I).

2. *There is moderate to severe anteversion* if the anteroposterior view shows apparent valgus and the frog-leg lateral shows considerable anterior angulation of the femoral neck (Fig. 5, II). In actual valgus the head usually sits higher than normal However, valgus cannot be diagnosed from an anteroposterior view alone. A long neck may be associated with severe anteversion (Fig 5, III).

3 *Anteversion probably is normal* if the lateral view (Fig. 5, IV) shows the head closer to axis of shaft than anteroposterior view.

4. *Retroversion is present* if the lateral view shows the head to be posterior to the axis of the femoral shaft (Fig. 5, V).

5. If both anteroposterior and frog-leg views (Fig. 5, VI) show a long neck with little angulation of the neck, the femoral neck is in *true valgus*, and any calculation of anteversion is probably of little significance.

## CLINICAL ESTIMATION

The value of physical examination in estimating anteversion must not be neglected. With the child sitting on the examining table with knees and thighs flexed to 90°, the range of internal and external rotation of the thighs (hips) is observed (Fig. 6). If internal rotation is 80° or more and external rotation is only 20° or less, moder-

FIG 5. (I) T. H., 3½ years Anteversion about 45°. Cervicofemoral angle about same in anteroposterior and lateral. (II) L. B , 3½ years. Severe anteversion (measured 62°) Anteroposterior. apparent valgus. Lateral: considerable posterior displacement of femoral shaft (III) B. B., 9 years. Severe anteversion. Anteroposterior: apparent valgus (IV) N. B., 5 years. Anteversion normal. Anteroposterior: cervicofemoral angle average. Lateral: slight posterior displacement. (V) S. M., 9 years Retroversion. Lateral shows anterior displacement of shaft. (VI) S B., 4½ years. Neck in true valgus.

ately severe anteversion is present, and the child probably walks pigeontoed, sleeps on the stomach with legs rotated internally, and sits on the floor between his legs. This clinical method of estimation is of less value in infants or in dislocated hips. Routine physical examination of a child should include checking the internal and the external rotation of the thighs.

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## Anteversion del Cervice Femoral; Un Methodo de Determination per Medio de un Triangulo Rectangule

*Summario in Interlingua*

Le grado de anteversion del cervice femoral pote esser mesurate directemente per medio de duo roentgenogrammas anteroposterior. In le prime le coxas se trova in position anatomic. Le secunde es un exposition lateral a "gambas de rana," i.e. (a) le genus es flectite a 90 grados, (b) le tibias es supportate parallel al tabula—in position

horizontal—sed il non es necessari que le tibias es parallel le un al altere, e (c) abduction es effectuate in tanto que le subjecto pote sequer sin disconforto. Iste position produce un rotation del femore per 90 grados pro le projection lateral.

Anteversion del cervice femoral o torsion femoral es un angulation anterior del capite e del cervice femoral in relation al axe transcondylar.

In altere parolas, si in le position anatomic del femore le diaphyse femoral es dosplaciate retrorsemente e etiam lateralmente in relation al capite femoral, on ha demonstrate le presentia de anteversion.

Le supra-describite position rende possibile mesurar in le roentgenogramma le duo distantias hic designate como "a" e "b," i.e. "a" = le displaciamento posterior del axe del diaphyse femoral ab le centro del capite e "b" = le correspondente displaciamento lateral. Un angulo triangule es construite in que le altitude e le base es proportional a "a" e "b." Le angulo "A" opposite al latere "a" es mesurate. Illo es le angulo del anteversion.

Attention: Non mesurar le false angulo! Si le diaphyse femoral monstra un displaciamento lateral in excesso del displaciamento posterior, le anteversion debe esser minus que 45 grados.

Angulo "A" pote etiam esser trovate secundo le formula:

$$\text{tangente } A = \frac{a}{b}$$

In consequentia del curvatura anterior del femore, un linea representante le axe del integre diaphyse femoral passa in le vista lateral a transverso le cortice posterior del tertio medial del diaphyse e le termino superior del diaphyse.

In juveniles, le centro del metaphyse pote esser considerate como le centro del capite.

Anteversion normal in adultos es inter 8 e 18 grados. In parvulos, anteversion normal pote attinger 50 grados.

Anteversion anormal es incontrate frequentemente in (a) congenite subluxation o dislocation del coxa, (b) paralyse partial, post poliomyelitis o paralyse cerebral, e (c) problemas de ambulatura, tanto a digito pedal extrorse como etiam introrse.

Familiaritate con iste methodo permitte al medico facer estimationes approximative del grado de anteversion exclusivemente super le base de un examine del mentionate expositiones special antero-posterior e a gambas de rana.

Si ambe expositiones monstra le capite a plus o minus le mesme distantia ab le axe del femore, le anteversion amonta a circa 45 grados.

Si le exposition antero-posterior revela valgo apparente durante que le exposition lateral a gambas de rana monstra grados considerable de apparente angulation anterior del cervice femoral, le anteversion es moderate o sever.

Si le exposition lateral monstra le capite in un position plus proxime al axe del diaphyse que le exposition antero-lateral, le anteversion es probabilemente normal.

Si tanto le exposition antero-lateral como etiam le exposition a gambas de rana monstra pauc angulation del cervice, le cervice femoral es in valgo e omne calculation del grado de anteversion es probabilemente sin grande signification.

Si in parvulos satis avantiate pro ambular le examine physic revela un excessive rotation interne insimul con un reducite rotation externe del diaphyse femoral, isto es usualmente un indication del presentia de anteversion. Assi un satis accurate estimation es obtenite.

# 9

# The Role of the Orthopaedic Surgeon in a Crippled Children's Program; Experiences in an Urban Community

ROBERT S. SIFFERT, M.D.*

The criteria used to measure the caliber of care of handicapped children and to set the goals for a crippled children's program are not necessarily similar in all communities. There are no absolutes that are applicable universally in evaluating services for orthopaedically handicapped children, whether rendered in hospitals, public health agencies, clinics or schools. In fact, the requirements of children in urban and in rural communities may vary tremendously because of basic differences in ways of life, as well as in vocational, social and educational standards. The technics of rendering health services themselves may vary also, depending on the availability of these services or differences in philosophy as to how they should be practiced Further, because of basic differences in the administration of state crippled children's programs, local communities of similar size with similar facilities may have completely different programs.

Remarkable changes have occurred in the nature of clinical orthopaedic practice during the last two decades. The most outstanding has been the decline in the incidence of chronic conditions, such as osteomyelitis, tuberculosis and poliomyelitis, because of antibiotic and prophylactic vaccine therapy, and public health measures. Teaching programs in medical schools, internships and residencies, as well as postgraduate education of the practicing physician through medical journals, graduate courses and inservice training programs for school-health and child-health physicians, have emphasized the importance of early diagnosis and early case-finding. When early referral is made, improved surgical and rehabilitation technics can be effective in *reducing the late deforming effects of trauma* as well as of congenital and developmental deformities that formerly required long and repeated periods of hospitalization. Changes in population trends, availability of hospital and ambulatory medical care, as well as educational, vocational and economic factors, have also been involved in the steady decline in the number of hospital-treated orthopaedic cases and the steady increase in numbers of those seeking and requiring ambulatory treatment Therefore, as orthopaedic surgeons, it may be well for us, in view of constant changes, to re-evaluate our role and responsibility periodically to ensure *that orthopaedically handicapped children* of the present and the future will receive the maximum benefit from ever-advancing skills of surgery and rehabilitation. Clinical ortho-

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paedic knowledge and experiences can serve administrators of hospitals and public health directors of state and local agencies as the most important baseline and guide in their current and future planning of realistic community programs for the care of these children. Through affiliation with local health agencies and local medical societies, the orthopaedic surgeon can participate and offer leadership in the guidance of crippled children's programs toward the development of comprehensive and co-ordinated care. When he does *not* participate, or offers an opinion in consultation only when solicited, there is no real guidance, and the emphasis tends to be more on the administrative than on the dynamic medical and total care aspects of the program.

The nature of this type of co-ordination must necessarily vary from community to community. In smaller areas, individual clinics or individual physicians may be the principal source of care for orthopaedically handicapped children. In the larger communities, care is centered more effectively within larger general institutions, where all medical (orthopaedics, pediatrics, physical medicine and rehabilitation, and other consultative services) and paramedical (nursing, social service, administration, vocation, education, recreation, etc.) facilities are available

Experiences with the New York City Crippled Children's Program (Medical Rehabilitation Program), administered through the Bureau for Handicapped Children of the Department of Health, with which the author has been associated for some years as consultant, form the basis of this discussion, which, in general, expresses the philosophy and the goal of comprehensive care for these children in this large urban community.

There is much diversity of opinion as to the role of a health department or a community health agency in a crippled children's program. Since public health agencies are charged with the responsibility of maintaining a high caliber of total health care in the community, they should also be concerned with the quality of care available to orthopaedically handicapped children. A community program cannot succeed if it is based on preconceived concepts. Careful and detailed fact-finding constitutes the *first step*. Before realistic plans can be evolved within a community, it is necessary to make personal surveys of local facilities, both clinical and nonclinical, in hospitals, clinics and treatment centers, as well as of the community facilities available, including education, social service, public health nursing, vocation and recreation. The surveys must include a *team* evaluation with representatives of all medical and paramedical specialists present, so that the information obtained will represent all areas of medical and paramedical care.

Once these facts have been collected, a philosophy of care must be evolved as the *second step*. The frame of reference used by the health agency in putting this philosophy into practice is actually a set of ideal *standards*—standards that both the institutions and the community agree to subscribe to in striving toward the development of a caliber of care worthy of the efforts and the carefully considered thinking of those concerned with the total well-being of the orthopaedically handicapped child. The connotation of *standards* often implies a rigid set of preconceived or beaurocratic principles. Therefore, it is of the utmost importance that all concerned have a crystal-clear picture as to concept. This perhaps should be defined rather as a *guide* than a set of standards, representing ideal goals that will be used as the blueprint for planning and improving total services within the institutions in the community and in the community as a whole. Basically, therefore, it must represent a frame of reference to which all concerned agree, including orthopaedists, pediatricians, physiatrists, public health physicians and administrators, other medical and paramedical specialists, institutions and commu-

nity agencies, so that efforts are directed uniformly and services are not duplicated.

For these reasons the *guide* never should be evolved without the co-operation of interested and concerned medical and community personnel, preferably in the form of an advisory committee. In smaller areas, medical, paramedical and community agencies may be consulted individually, but, where possible, it is generally more effective to develop a formal advisory committee, preferably with rotating membership, that is widely representative of medical and community leaders from appropriate medical and paramedical specialties By this technic, not only can the orthopaedist obtain support for his program, but he can benefit from the experience and the opinions of those concerned in other facets of care of the orthopaedically handicapped child.

The fact that a set of standards has been developed should not be accepted as the final answer. As clinical practice changes and hospital consultants and hospital staffs acquire further knowledge and understanding, periodic revision of established standards is indicated. Too often the standards adhere to a bygone era.

These first two steps, i.e., fact-finding and goal, or *standard-setting,* acquire practical significance only when they become part of an ongoing dynamic program under the guidance of the local health agency. This depends upon close relationships among all participating physicians, institutions and agencies, and also upon frequent conferences of the health agency, the advisory committee and the participating institutions and physicians to discuss the philosophic and the practical direction of future planning. Therefore, all institutions benefit from the pooled experiences of other institutions in the community faced with similar problems. The orthopaedist must be able to interpret to the medical and the paramedical personnel the suggestions and the recommendations of the advisory committee, gearing them specifically to the requirements of the handicapped children in that community, as well as to the limitations of and the facilities available in the particular institution. To overburden an institution with idealistic and impractical goals is frustrating and confusing, but sensible and realistic step-by-step recommendations possible of accomplishment will be helpful. Therefore, an ongoing program consists of periodic re-evaluation of an institution's needs, problems and goals in reference to those of the community as a whole; conferences with the advisory committee productive of realistic recommendations to the institutions and the health agency; and reappraisal of the role of the various institutions in the light of medical and community changes that eventually may necessitate reconsideration of the goals of the crippled children's program as a whole.

Important as improving existing facilities may be, the influencing of administrators of programs toward expansion should be considered. Examples demonstrating the fact that handicapped children are not reaching their optimum potential because of lack of therapy or vocational, educational or other community facilities or opportunities usually open new vistas in public health planning in the development of these services. An advisory committee of outstanding community leaders, convinced of these needs, can be of enormous help in influencing public health and civil administrators. Usually, not until a need is demonstrated medically are steps taken in the community to cope with it. For example, it is the orthopaedist who can document the change in needs for better outpatient facilities with more inclusive services; for better teaching programs for pediatricians and general practitioners in early diagnosis; for better follow-up technics and better facilities for education, vocational training and rehabilitation of the handicapped child. By insisting upon the importance of early diagnosis in preventing permanent crippling deformities, he alone may convince the directors of pediatrics to include orthopaedic outpatient training in their

residencies so that they may become familiar with *office* orthopaedics, hospital administrators to expand clinic facilities, and public health administrators to expand community resources and vocational and educational opportunities.

In New York City, qualified orthopaedic surgeons who are engaged in private practice and are active members of hospital staffs serve as part-time consultants on the crippled children's program. Each has a responsibility for a particular part of the program, participating actively in both the current and the future planning for better care. Their various roles are outlined briefly as follows.

## HOSPITAL CONSULTATION

Because of an awareness that its community responsibility toward orthopaedically handicapped children should extend beyond the administrative role and payment for care, the Health Department has been concerned that (1) the over-all care being rendered should be of good quality; (2) every assistance possible should be offered to improve the caliber of existing care; and (3) new services should be planned in anticipation of future medical and community needs.

In the area of fact-finding, hospitals participating in the crippled children's program were surveyed personally by a team consisting of consulting specialists in orthopaedics, physical medicine and rehabilitation, and pediatrics, a public health nurse and a medical social worker of the Bureau for Handicapped Children At the hospital the personnel in these fields were met, the care of the children was discussed, and the inpatient and the outpatient facilities were surveyed.

The Orthopaedic Advisory Committee, which was under the chairmanship of Dr. Philip D Wilson for many years, is composed of 21 community leaders in medical and paramedical specialties. The committee established a set of *standards* to be used as the blueprint, or goal, of the program. Using the standards as a guide, the Orthopaedic Advisory Committee made specific and realistic recommendations for institutions. The survey team returned to the institution, explaining that the recommendations being discussed and clarified were not to be regarded as criticism but as suggestions based on the experience of a large number of surveys of similar institutions as well as of the Advisory Committee's experiences in the area of good total patient care. The conference was attended by the same medical and paramedical personnel who were at the original survey, including the hospital administrator and his staff. The hospital staffs, many members of which had known each other only through administrative relationships, often had their first opportunity to discuss the common problems pertaining to the care of orthopaedically handicapped children and to observe the co-ordinated team thinking of the Health Department consultants in discussing comprehensive care. Frequently, following the conference, plans were made for continued contact between the Health Department and the hospital to follow up specific suggestions and plans. By periodic contact with the institution, ranging from conferences to annual complete surveys, depending on the problems involved, or the institution's desire for the assistance of the hospital consultation team, hospital consultation has remained an important part of the program of the Bureau for Handicapped Children and has resulted in much improvement in care over the last 8 years.

Because of the marked fall in inpatient figures in New York City and the greater need for outpatient facilities, hospital consultation during the last 2 years has concentrated more on outpatient care. Using the successful technics outlined above, the outpatient departments of the 20 participating hospitals have been surveyed personally by a team, and the observations have formed the basis of a set of *standards*, or *guide*, for ambulatory care similar to that developed for inpatient care. With this as a baseline, the team now is revisiting the hos-

pitals for conferences in first steps designed to improve ambulatory care of the orthopaedically handicapped children in New York.

## REVIEW COMMITTEE

Once a hospital has been approved for participation in the Medical Rehabilitation Program, the technical orthopaedic care rendered becomes the sole responsibility of the medical staff of that institution. Administrative problems always arise relating to eligibility and time limitations of care, making it necessary that there be a certain amount of *paper review* of applications by an orthopaedist. He raises questions with the institutions and in most instances can manage the problems. Complicated and involved situations are laid before a review committee that meets periodically and consists of the Bureau director, 2 orthopaedic consultants, a pediatric consultant, a rehabilitation consultant, a public health nurse and a medical social worker.

The Review Committee was begun in 1952 because hospital surveys revealed a large amount of overstay and overinstitu*tionalization. Analysis showed that some* of the reasons for this overinstitutionalization were inadequate medical supervision of patients, infrequent team evaluations of patients, too little and too late social service coverage of the patient and his family, unfamiliarity with community resources, and gaps in resources in the community to meet the needs of handicapped children. It was felt that in many of these areas the Health Department could be of help, and records of children in institutions for more than 3 months were reviewed regularly. By offering assistance, overinstitutionalization was greatly reduced. In the process, hospitals became aware of the problems in these areas of community resources and of the need for early social service planning. Review of the records acquainted the Health Department team with specific problems within specific hospitals that later became the subject of discussion and co-operative efforts at future hospital surveys and conferences. As a result of careful and intensive work in this direction, the need for close scrutiny no longer exists, and most problems can now be handled by the reviewing orthopaedist alone. The Review Committee has become a team conference, meeting regularly with the Bureau director to review difficult cases and problems in general, where the combined thinking of the consultants in the various fields forms the basis for future planning of services, defining of policies and activities of the Bureau.

## ORTHOPAEDIC CONSULTATION SERVICE

Although most children are referred to hospitals and clinics for orthopaedic consultation, the Health Department lends additional diagnostic support to case-finding agencies, such as school health, child health stations, employment centers, and physicians who may have patients unable to afford this type of service privately. The service, for which there is no charge, is located physically in a major hospital and staffed by a *Health Department orthopaedic consultant,* a public health nurse and a medical clerk. The hospital setting was chosen rather than a local Health Department clinic because it offers x-ray and laboratory facilities. At a single visit these services are performed, as indicated, to arrive at a diagnosis, and a consultation report is sent to the referring physician with recommendations for further management. It is suggested that if further orthopaedic attention is needed, the referring physician send the child to an orthopaedic surgeon or to an institution approved under the Medical Rehabilitation Program. The Orthopaedic Consultation Service has been a source of early case-finding, serving as an educational experience by offering physicians personal consultation in case-finding agencies so that they may follow the patients whom they refer for opinion and actually participate in their continued man-

agement. More than half of the children seen have been found to be in need of active orthopaedic treatment that often might not have been rendered if this method of easy and personal referral for consultation were not available. Many times the orthopaedic surgeon's evaluation assists materially in the appropriate educational placement of the child, either in a special class, an orthopaedic class, a school for the handicapped or home instruction.

## SCHOOL AND CHILD HEALTH SERVICES

The orthopaedic consultants co-operate with school and child health services in teaching, in school placement and in medical supervision in schools of orthopaedically handicapped children. An orthopaedic consultant participates in inservice training programs for physicians in school and child health services, familiarizing them with clinical orthopaedic problems and methods of diagnosis in an attempt to improve early case-finding. Members of the orthopaedic consulting staff review records and recommendations for school placement and consult with private physicians in an attempt to guide children to as normal a school environment as possible compatible with their disabilities. The orthopaedic surgeons participate in care at 5 special school units to which severely handicapped cerebral palsy and orthopaedically handicapped children are transported by bus and receive education and physical, speech and other indicated therapies as prescribed by their treatment agencies.

Another team, consisting of an orthopaedist, a public health nurse and a medical social worker, reviews recommendations for home instruction. In those situations in which there is the possibility of some school placement, further investigation by the public health nurse and contact with the treating agency, if one exists, often uncover neglected children with medical, social or nursing problems who need assistance. The design is to get as many children as possible into as normal a school environment as possible, and it has been a further source of case-finding throughout the city.

## OTHER ROLES

Further areas in which orthopaedic surgeons play a role in New York City's Crippled Children's Program are: consultation and conference with other bureaus in the Health Department, such as Public Health Nursing and Child Health, and with the Department of Education as well as community agencies involved in child care; participation in Health Department conferences, seminars, radio broadcasts, conferences with health departments of the state and other communities, medical society presentations and conferences, and new programs, such as establishing standards for cerebral palsy and amputee centers.

The possibility of clinical research is now being investigated. A number of orthopaedic conditions capable of roentgenographic evaluation are of relatively infrequent occurrence in the practice of a single orthopaedic surgeon or in the experiences of a given hospital. For example, Perthes' disease lends itself to radiologic evaluation of large numbers of cases treated in statistically significant numbers by each of the various methods currently practiced (ranging from no treatment to braces, slings and complete bedrest). This study, carried out by collecting roentgenograms through photographic technics and assembling clinical data from records, is currently in progress, and other clinical research projects are being planned.

There are countless ways in which the orthopaedic surgeon can participate in community programs for improvement of care of orthopaedically handicapped children. The drawback is the relatively small amount of time that the busy orthopaedic surgeon can devote to his community in applying his skills and knowledge to the children who

*require it. His value to the community and* to the health agency in community planning is in direct proportion to his expertness as an orthopaedic surgeon, a fact alone demanding that the greater portion of his time be devoted to the practice of clinical orthopaedic surgery. *Because of this limitation of* time, as many orthopaedic surgeons as can possibly participate in the program should do so, for on their availability, interest, enthusiasm, ability, devotion and ingenuity depends the success of the program.

## Le Rolo del Chirurgo Orthopedic in un Programma pro Infantes Deforme; Experientias in un Communitate Urban

*Summario in Interlingua*

Le experientias del autor como consulente orthopedic in le Programma pro Infantes Deforme de un grande communitate urban (in le citate de New York) es usate como base de un discussion del methodos per le quales un practicante chirurgo orthopedic pote contribuer al melioration del cautela total del orthopedicamente handicappate infantes de su communitate. Le prime passos necessari es establir le factos con respecto al facilitates disponibile intra le communitate e le formulation, al uso del communitate, de un corpore de "standards," i.e. de un un "guida" verso le "objectivo" que on vole attinger. Le ideas subjacente un Programma pro Infantes Deforme deberea esser le resultato de effortios coordinate del parte del administratores del hygiene public e de chirurgos orthopedic, e etiam de altere specialistas medical (i.e. de pediatria, medicina physic, e rehabilitation) e paramedical (i.e. de infirmeria, servicio social, direction vocational, education, etc.). Gratias a su cognoscentias clinic, le chirurgo orthopedic es qualificate a ager efficace- e dynamicamente como orientator in le effortios a meliorar le cautela currente e a planar programmas futur de maniera que le infantes obtene le maximo possibile de beneficio ab su capacitates chirurgic e rehabilitational.

# 10

# Osteomyelitis Since the Advent of Antibiotics; A Study of Infants and Children*

GORDON M. COTTINGTON, M.D., JAY M. RIDEN, M.D., AND ALBERT B. FERGUSON, JR., M.D.†

Osteomyelitis of infants and children is a disease that has caused chronic drainage, deformities and death. In reviewing a series of cases in infants and children, certain differences between the two age groups have been noted. Antibiotics have lowered the number of deaths but have not greatly changed the chronicity or the deformities produced by this disease in infants. In the older-age group, often the use of antibiotics has produced a low-grade smoldering bone abscess. The incidence of the disease from the standpoint of admissions to Children's Hospital has remained very much the same as that of the preantibiotic era. The diagnosis of osteomyelitis can be made from the roentgenogram early in the course of the disease, and the complications are preventable if proper surgical measures are taken at the onset of the disease. Osteomyelitis is still a surgical emergency, and its complications cannot be eradicated by medical treatment alone.

## PAST HISTORY

John Homans,[6] writing in 1912, reported 94 cases of osteomyelitis of the long bone from the Children's Hospital, Boston, over an 8-year period. Slightly less than 10 per cent died, and almost 20 per cent healed with deformity. This involved all childhood age groups.

Green and Shannon,[5] writing for the same hospital in 1936, reported 95 cases below the age of 2 within 21 years. The mortality in infants was 22 per cent with 45 per cent mortality under 6 months of age. Fifty-four of these cases were on the orthopaedic service, where 46 survived. Of these, 5 were left with residual deformity from joint sepsis and 5 from partial destruction of the epiphysis of a growing bone. Recurrences were rare, healing was rapid. Streptococcus was the most prominent organism under 2, with staphylococcus being more common over this age.

Blanche,[3] in 1950, reported 35 infants in 16 years from Los Angeles Children's Hospital. The youngest was 14 days old; the oldest, 9½ months. Staphylococcus constituted the major organism. Of 20 cases involving the femur, 16 were at the proximal end. Eleven of these involved the hip. Eight of these hips were already dislocated when first seen. There were 2 deaths, and 44 per cent were left with a major deformity of a crippling nature. The disability ap-

* Aided by a grant from the Western Pennsylvania Chapter of the Arthritis and Rheumatism Foundation.

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require it. His value to the community and to the health agency in community planning is in direct proportion to his expertness as an orthopaedic surgeon, a fact alone demanding that the greater portion of his time be devoted to the practice of clinical orthopaedic surgery. Because of this limitation of time, as many orthopaedic surgeons as can possibly participate in the program should do so, for on their availability, interest, enthusiasm, ability, devotion and ingenuity depends the success of the program.

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FIG. 1. Destruction of the epiphysis by metaphyseal osteomyelitis is a common cause of deformity from childhood bone and joint diseases.



FIG. 2. Septic dislocation of the hip with loss of head and neck secondary to joint infection.

damage was noted on the first roentgenogram at the time that the diagnosis was first made. None developed after treatment was instituted. From these figures it is obvious that earlier diagnosis is the key to the prevention of development of complications leading to a crippled child, and the physician cannot feel secure merely because the child is getting chemotherapy.

The clinical diagnosis will not be overlooked if it is remembered that these children can be quite ill and harbor bone and joint infections without a marked systemic reaction Swelling of the extremity appears early in infancy and childhood, and may be quite diffuse. Failure to move the extremity even to the point of apparent flaccid paralysis is the most common presenting complaint. The most valuable sign allowing recognition of the septic hip is the determination of a flexion contracture. The usual flexion contracture at the hip with sepsis involvement is at least from 60° to 70°.

## ROENTGENOGRAM

Osteomyelitis can be diagnosed in the first few days of onset; it is not necessary to wait for visible bone damage to make the diagnosis. The soft-tissue changes visible in the roentgenogram in the presence of osteomyelitis must be read accurately. Osteomyelitis is accompanied by a deep soft-tissue swelling centering about the metaphysis. This is readily recognized and differentiated from swelling present in cellulitis, which involves the subcutaneous tissue predominantly. Osteomyelitis always originates in the metaphyseal area, although it may track into the joint or down the shaft.

The deep swelling has poorly defined

peared to be directly proportional to the delay in instituting treatment. It was quite usual for these infants to have a temperature below 100° and a white count not particularly elevated for this age group at the onset of the disease.

## CHILDREN'S HOSPITAL OF PITTSBURGH SERIES

### UNDER 3 MONTHS

Cases recorded at the Children's Hospital of Pittsburgh were reviewed. The time interval for infants spans 1940 to 1957. There were 36 infants, all under 3 months of age, and over half were first diagnosed from the eleventh to the thirteenth day. Only 6 were diagnosed in the first 10 days of life. Streptococci and staphylococci were equally divided as organisms. The pneumococcus has almost disappeared as a cause of osteomyelitis.

There were 3 deaths: 1 in 1941 and 1 in 1942, both treated with sulfathiazole and Dakin's tube drainage; and the third in 1955, a septic hip that was treated with aspiration only. A mortality of 7 per cent was noted.

There were 9 septic hips, 5 of which were shown to be dislocated on the first roentgenogram taken. Four were not dislocated, and under treatment they remained undislocated. The very definite implication that femoral vein puncture was an etiologic factor in the localization of infection in the hip joint of 4 of these youngsters is of definite interest and should rule out the femoral vein as a blood source in septicemic infants.

### OVER 3 MONTHS

In children over the age of 3 months, a survey was made from 1928 to 1956. It comprised 154 cases. The most frequent sites of osteomyelitis over 3 months of age were (1) multiple sites, (2) the distal femur, (3) the proximal tibia and (4) the hip, in contrast with the infant group in which the hip was most commonly involved, the other sites then following in the same order

Septicemia was most common as the precipitating factor, with soft-tissue abscesses and upper respiratory infections next in frequency.

Hemolytic *Staphylococcus aureus* was the organism cultured in 90 per cent of cases in children, in contrast with the nearly equal ratio of staphylococcus and streptococcus in infants.

The mortality rate was 6.4 per cent. When diagnosis was delayed, septic hips resulting in dislocations later required major reconstructive surgery.

Chronic drainage was noted to increase sharply with the greater delay in onset of treatment, and, even in cases seen 3 to 7 days after onset of symptoms, 50 per cent developed chronic drainage. Surgical treatment here included both inadequate and adequate measures. In cases treated with antibiotics, 46 per cent continued to have chronic drainage with deformities, whereas in those prior to the antibiotic era, 76 per cent had chronic drainage and deformities. While this is a reduction of 30 per cent, still virtually half of the cases continued to have chronic drainage and deformity and required surgical correction.

## DISCUSSION

The concept that chemotherapy has eliminated the recurrence of bone infection in those who had an onset in infancy does not stand up as important when it is remembered that recurrences were rare before chemotherapy in this age group. Recurrences are rare in infants because of the tendency to decompress spontaneously through the relatively spongy vascular bone and the thin cortex.

The mortality appears to be improved and is presumably due to the advent of chemotherapy.

Major crippling deformity involved 10 per cent in all age groups in 1912, 20 per cent under 2 in 1956, 44 per cent under 9½ months in 1950, and 29 per cent under 3 months in 1957. In all the cases with crippling deformity in the last two series, the



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The clinical diagnosis *will not* be overlooked if it is remembered that these children can be quite ill and harbor bone and joint infections without a marked systemic reaction. Swelling of the extremity appears early in infancy and childhood, and may be quite diffuse. Failure to move the extremity even to the point of apparent flaccid paralysis is the most common presenting complaint. The most valuable sign allowing recognition of the septic hip is the determination of a flexion contracture. The usual flexion contracture at the hip with sepsis involvement is at least from 60° to 70°.

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The deep swelling has poorly [illegible]

edges and involves the so-called muscle shadow of water density. Fat lines seen normally in the muscle shadows are obliterated by the inflammatory changes. The septic hip itelf is recognized by swelling of the capsular shadows about the joint and widening of the joint recognized at its inferior medial aspect.

## TREATMENT

In order to avoid the development of hip dislocation due to sepsis, it is necessary to institute surgical drainage of a tense, pus-filled hip joint as soon as the diagnosis is certain. A preliminary needle aspiration is done, and, if pus is obtained, the patient is turned face downward, and drainage is accomplished through a posterior incision.

It is the usual procedure to drain subperiosteal abscesses when they are clearly evident and fluctuation is apparent, or when there is any possibility of involvement of the epiphyseal line. Drainage is not done for cellulitis alone in infants. Drainage should be performed for osteomyelitis in children as soon as the diagnosis is made, and this is done by an evaluation of the clinical picture and the soft-tissue roentgenographic changes. The bone is drained with drill holes made through the cortex *without elevating the periosteum* to ensure adequate decompression of the medullary cavity. It is important to maintain the blood supply to bone in order to avoid the development of sequestra secondary to surgery.

## SUMMARY

The use of joint and abscess drainage appears to be adequate to prevent the development of crippling deformity provided that the damage has not already been done. Osteomyelitis in infants is different from sepsis in the older-age groups. The natural anatomic and physiologic characteristics of infants are in large part so much on the side of the physician that complications can be avoided with proper treatment. Antibiotics have lowered the mortality rate but have not prevented the biggest single complication—major crippling deformity. Intelligent use of surgery is still necessary to prevent the destruction of tissue and consequent development of deformity attendant upon infection of the bones and joints in the growing child.

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## Osteomyelitis Depost le Advento del Antibioticos; Un Studio del Morbo in Infantes e Juveniles

*Summario in Interlingua*

Osteomyelitis in infantes e juveniles ha essite le causa de drainage chronic, de deformitates, e de morte. In juveniles de un etate plus avantiate, le uso de antibioticos ha frequentemente producite abscessos ostee a "combustion surrepticie." Le incidentia de deformitates invalidante in consequentia del morbo ha remanite, al Hospital Juvenil de

Pittsburgh, sin alteration significative in comparation con le situation pre-antibiotic.

Esseva trovate que septicemia es le plus commun factor precipitatori. Puncturas del vena femoral esseva clarmente implicate in quatro inter nove casos de coxa septic in infantes.

Hemolytic *Staphylococcus aureus* esseva culturate in novanta pro cento del patientes de plus que tres menses de etate. Staphylococcos e streptococcos esseva culturate in equal procentages in le patientes de minus que tres menses de etate.

Esseva constatate un acute augmento del incidentia de drainage chronic in casos in que le tractamento habeva essite retardate. Cinquanta pro cento del casos continuava haber drainage chronic in despecto del tractamento antibiotic.

Un plus prompte establimento del correcte diagnose super le base de examines clinic e de alterationes trovate in le radiogrammas es un desiderato urgente in le interesse del prevention de chronicitate e de deformitates invalidante

Osteomyelitis pote esser diagnosticate per medios radiographic intra pauc dies post su declaration. Illo se manifesta in le apparition de tumescentia del profunde histos molle circa le metaphyse. Pro facer le diagnose, le medico non debe attender visibile lesiones ossee in le radiogramma.

Un tense articulation que es replenate de pus debe esser tractate per drainage chirurgic post le effectuation preliminari de aspiration a agulia. Isto es un mesura de urgentia que es necessari pro prevenir le dislocation del coxa. Il es costumari drainar abscessos subperiostee si tales es presente. In infantes, cellulitis per se non indica drainage. Le osso es drainate per medio de forationes que transversa le cortice sin elevar le periosteo. Le objectivo de isto es preservar le alimentation de sanguine e evitar le formation de sequestrationes.

Le effectuation de drainage de articulation e abscesso, si interprendite satis precocemente, preveni deformitates invalidante, providite que le natural characteristicas anatomic e physiologic del infante permitte le evitation de complicationes per medio del appropriate tractamento. Le uso de antibioticos ha reducite le mortalitate, sed illo ha non potite prevenir le major deformitates invalidante. Le uso intelligente de mesuras chirurgic remane indispensabile pro prevenir deformitates in consequentia de osteomyelitis in juveniles crescente.

# 11

# Engelmann's Disease (Progressive Diaphyseal Dysplasia)— A Nonprogressive Familial Form of Muscular Dystrophy With Characteristic Bone Changes

BERTRAM R. GIRDANY, M.D.*

Engelmann's disease is a relatively rare affection of the skeleton often associated with muscular weakness. The diagnosis derives from characteristic roentgen changes in the skeleton described by Engelmann[3] in 1929. Evidence is presented here that the condition may be familial, that the associated muscular weakness is not necessarily progressive, and that the typical bone changes may be present in asymptomatic individuals.

## LITERATURE

Twenty-five case reports of Engelmann's disease have appeared in the literature. Cockayne,[2] in 1920, reported diaphyseal sclerosis in the long bones of a 9½-year-old male. Camurati,[1] in 1922, reported sclerotic changes in the bones of the lower extremities of a 55-year-old father and his 7-year-old son. Engelmann[3] reported changes in the skeleton of an 8-year-old white boy who had had pains in his legs since the age of 4. The child was poorly developed and walked with a peculiar *wobbly* gait. Ortolani and Castagnari, in 1933, recorded bone changes in a 6-year-old girl who complained of easy fatiguability and muscular weakness. Roentgenographic examination 20 years later showed no progression of the sclerotic changes.[7] Her asymptomatic 19-year-old sister showed diaphyseal cortical sclerosis in the bones of her lower extremities. Riley and Shwachman,[9] in 1943, reported diaphyseal sclerosis in the skeleton of a 4¾-year-old white girl whose complaints were weakness, faulty gait and malnutrition. Neuhauser,[6] in 1948, described skeletal changes in 3 children and applied the term *progressive diaphyseal dysplasia.* Stronge and McDowell,[10] Griffiths,[4] and Mikity and Jacobson[5] recorded bone changes of Engelmann's disease in a 28-year-old man, a 29-year-old woman and a 54-year-old man, respectively, none of whom had major complaints referable to the neuromuscular system. Ribbing[8] reported diaphyseal sclerosis in some of the

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long bones of 4 members of the same family. He called these changes *hereditary multiple diaphyseal sclerosis* and did not consider them to be examples of Engelmann's disease.

Eleven biopsies have been recorded. All specimens showed thickening of the cortex produced by both endosteally and subperiosteally accumulated new bone. Abnormal cells were not present.

## CASE REPORTS

The author has observed 12 adults and children with Engelmann's disease. In 10, all the long bones and the skull were involved; in 2, the changes were confined to the tibias.

Case 1. A 3½-year-old white male was referred to the Children's Hospital of Pittsburgh because he tired easily, had a waddling gait and complained of muscle pain on exertion. He *climbed his frame* to assume a sitting position. His poor musculature led to a tentative diagnosis of atrophic type of progressive muscular dystrophy. Roentgenographic examination showed involvement of all the tubular bones (Fig. 1) and sclerosis of the base of his skull The child was placed on a regular schedule of physiotherapy; he was advised to avoid prolonged bedrest. Five years later his muscle weakness was less pronounced, and he was able to stand from a lying position normally (Fig. 2).

His 1½-year-old sister showed small areas of cortical thickening affecting the mid-portions of her long bones, the changes barely exceeding limits of normal. Five years later she walked with a waddling gait; the muscle masses in her extremities were small Roentgenographic examination revealed the findings of *progressive diaphyseal dysplasia* in her tubular bones and in her skull.

A maternal uncle, at age 14, was too weak to walk to school. Of his own accord he undertook a series of *dynamic tension* setting-up exercises; at age 21 he earned his livelihood by performing manual labor in a steel mill. His entire skeleton showed roentgenographic changes of Engelmann's disease (Fig 3). His musculature was normal.

The boy's 33-year-old mother had similar findings in all her long bones and skull. She had normal musculature and had no complaints referable to the neuromuscular system. A second maternal uncle, aged 30, whose roentgenographic examination demonstrated diaphyseal sclerosis in both tibias, was asymptomatic. His maternal grandmother, aged 57, showed the changes of Engelmann's disease throughout her skeleton (Fig. 4). There was evidence of freshly deposited subperiosteal new bone in this asymptomatic individual.

Case 2. The roentgenographic changes of *progressive diaphyseal dysplasia* in the long bones and skull were observed as incidental findings in a well-developed 23-year-old Negro woman who had a routine roentgenogram taken of her abdomen.

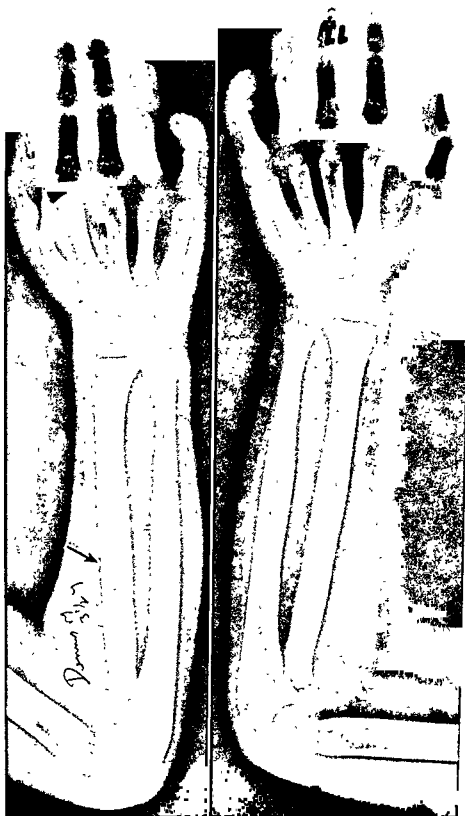
Case 3. All the long bones, ribs, spine and skull of a 10-year-old white boy, studied because of severe mental retardation, showed changes of Engelmann's disease. Neuromuscular evaluation of this child was not satisfactory.

Case 4. A 5-year-old white boy was examined because he had a peculiar gait and poor musculature. Roentgenographic examination of his skeleton showed changes of Engelmann's disease. Similar changes affected the entire skeleton of his thin 9-year-old sister. Diaphyseal sclerosis was present in the tibias of the father of these 2 children. The base of his skull was sclerotic.

Case 5. A 10-year-old poorly muscled, weak, colored girl, hospitalized for study because of tiredness, inability to walk and pains in her legs, showed changes of progressive diaphyseal dysplasia affecting all her tubular bones, skull and mandible (Fig 5). She had been examined first at 2½ years of age because of peculiar gait and because she cried when going down stairs. She had been treated f[illegible] *syphilis* at age 4, [illegible]er shins, and the [illegible]ving been diag- [illegible]riostitis. Muscle and tibial biopsies were done, and she was begun on a regular course of physiotherapy. At age 16 her general strength has increased, and she is able to walk long distances without tiring. Puberty has not been associated with any new muscular achievements; there has been some increase in the size of her gastrocnemius muscles

## DISCUSSION

The roentgenographic changes in Engelmann's disease consist of both internal and external cortical thickening confined to the diaphyses of the tubular bones. The metaphyses are not involved, and normal tubu-



FIGS. 1 to 4, D. M., a white male. FIG. 1. Roentgenograms at 3½ years of age of forearms and hands (*left*) and pelvis and femurs (*on facing page*) showing internal and external cortical thickening affecting the shafts of the tubular bones The proximal portions of both femurs are not affected, and the pelvis is normal.

lation is present. The earliest changes occur in the mid-portions of the shafts; the cortical involvement progresses to involve the major portions of the shafts. Abnormal deposition of cortical bone continues throughout life, the process acting as if there were an imbalance between the deposition and the absorption of cortical bone.

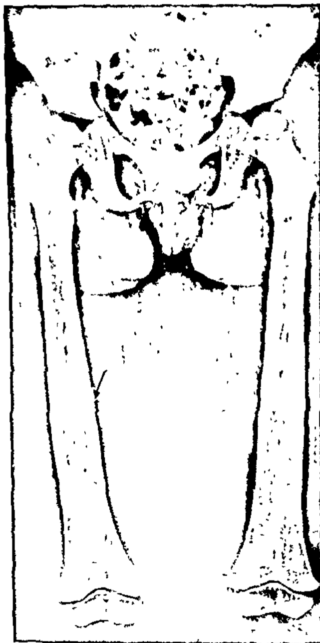




Fig. 1 (*Continued*). (*Caption on facing page*)

The base and the vault of the skull and the vertebral bodies with their pedicles may show sclerosis; the mandible occasionally shows marked cortical thickening (Fig. 6).

All children observed with this condition have been weak, have had small muscle masses and have walked abnormally. The bone changes have been present in asymptomatic well-muscled adults. Cortical sclerosis affecting single bones has been observed in siblings or parents of patients with universal changes of Engelmann's disease.

Information about the incidence and the course of Engelmann's disease is limited. The author's 12 cases indicate that the disease is not as rare as the small number of reports in the literature would suggest.

There are marked variations in the course of the disease. The bone changes may exist without muscle symptoms and be recognized as incidental findings on roentgenographic

genographic examination of the skeleton.

The condition may resemble other forms of nonpseudohypertrophic muscular dystrophy with history of progressive weakness. The response to physiotherapy and exercise suggests that the muscular weakness is not necessarily progressive. Avoidance of prolonged bedrest is a cardinal feature in the treatment of this disease, as it may possibly be in the treatment of all forms of muscular dystrophy. The effect of puberty on the progression of muscular symptoms is not established.

The differential diagnosis of Engelmann's disease includes all forms of nonhypertrophic muscular dystrophy. Roentgenographic examination permits definitive differentiation.



FIG. 2. Roentgenograms of the lower legs (*left*) and the left upper extremity (*on facing page*) at 6 years of age show the small muscle mass in the calves. The sclerosis has extended to involve major portions of the shafts of the bones Normal tubulation is present.

The bone changes may resemble Paget's disease but are not associated with abnormal chemical values in the ... uma, hypervitaminosis A and inf... hyperostosis are other causes ... ing of the tubular bones

It is probable that the ... by Ribbing[8] represent ... mann's ... The ... involv... skeleton of a family whose mother, siblings, niece and nephew showed universal changes and the observation of tibial sclerosis in the father of 2 children with Engelmann's disease support this presumption. Identification of diaphyseal sclerosis affecting individual bones as belonging to the Engelmann group depends on normal chemical values of the blood and the finding of similar changes in the skeleton of other members of the family.

## SUMMARY AND CONCLUSIONS

1. Twelve cases of Engelmann's disease are reported. Six of these occurred in 1 family with 3 generations affected. Two of the 12 patients were Negro females.

2. Muscular weakness is not necessarily progressive. Increase in general strength followed physiotherapy in 3 patients.

3. Roentgenographic examination of the skeleton is indicated in all patients with nonhypertrophic forms of muscular dystrophy.

4. The roentgenographic changes of Engelmann's disease may be present in asymptomatic individuals.

5. Involvement of single bones may occur as a variant of progressive diaphyseal dysplasia.

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FIG. 2 (*Continued*). (*Caption on facing page*)



FIG 3 (*Bottom*). Roentgenogram of the right upper extremity of the 21-year-old uncle of D. M. shows sclerosis of the shafts of the humerus and the radius. All his tubular bones were affected.



FIG. 4. Roentgenographic examination of the lower leg of the 57-year-old grandmother of D. M. shows the characteristic changes of progressive diaphyseal dysplasia. The muscle mass in the calf is normal, and there is evidence of freshly deposited subperiosteal new bone along the shaft of the tibia.

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## Morbo de Engelmann (Progressive Dysplasia Diaphysee)— Un Non-Progressive Forma Familial de Dystrophia Muscular con Characteristic Alterationes Ossee

*Summario in Interlingua*

1. In 1929 Engelmann describeva le caso de un patiente pediatric con *debilitate* muscular, exhibiente characteristic alterationes roentgenographic del skeleto. Depost ille tempore, al minus 25 simile patientes ha essite reportate in le litteratura. Le prognose

es considerate como nonfavorabile, e le termino "progressive" dysplasia diaphysee es usate pro iste condition.

2. Le presente articulo reporta 12 casos de morbo de Engelmann. Sex de illos occurreva in un sol familia, afficiente 3 generationes. Duo del 12 patientes esseva feminas negre.

3 Debilitate muscular non es necessarimente progressive. In 3 del patientes, physiotherapia esseva sequite per un augmento del fortia general.

4. Examines roentgenographic del skeleto es indicate in omne patientes con formas non-hypertrophic de dystrophia muscular.

5. Le alterationes roentgenographic de morbo de Engelmann pote esser presente in individuos asymptomatic.

6. Il existe un variante de progressive dysplasia diaphysee in que solmente ossos individual es afficite.



FIG. 5, M. O. Lateral roentgenogram of the skull shows the marked sclerosis of the base and thickening of the mandible.

# 12

# Spina Bifida Occulta in Legg-Calvé-Perthes Disease

Jacob F. Katz, M.D.*

In the course of analysis of the results of therapy in Legg-Calvé-Perthes disease, roentgenograms were reviewed in a large series of cases. Casual inspection of the lumbosacral segment included in the roentgenograms revealed a surprisingly high incidence of spina bifida occulta. This stimulated more careful search and study in order to explore a possible relationship between Legg-Calvé-Perthes disease and spina bifida occulta.

## HISTORICAL BACKGROUND

In past years spina bifida occulta has been the focus of attention intermittently by investigators with varied interests Much valuable information has come from studies of enuresis, the relationship of which to spina bifida occulta had long been accepted.

In 1922, Hintze[5] performed a convincing and meticulous study in which he found an average incidence of 67 per cent of spina bifida occulta in enuretics between 3 and 17 years. He did a companion study of 117 normal children between the third and the fifteenth years, in whom he found 55 per cent with spina bifida occulta. In a breakdown of the cases by age groups, the 3- to 5-year age span showed the highest incidence (90% in enuretics, 81% in normals) and the 15- to 17-year age span the lowest incidence (42% in enuretics) of spina bifida occulta. He concluded that spina bifida occulta did not differ in frequency in enuretic children and normal children, and that, because of its decreasing incidence with age, it was a normal condition in children.

Karlin[7] studied the problem in much the same way in 1935. He found spina bifida occulta in 84 per cent of 25 children who had enuresis; they were in the age group 3 to 14 years. In 50 control children, 54 per cent were reported to show spina bifida occulta. He concluded that spina bifida occulta was a developmental defect indicating some abnormality in the process of embryogenesis in the region affected and was of questionable relationship to enuresis.

Other workers attempted to relate orthopaedic anomalies and abnormalities to spina bifida occulta. Ingraham and Lowrey[6] studied 65 children with spina bifida occulta, 31 of whom presented extremity maldevelopment and gait disturbances. No special analysis of age-group incidence was done. They conceded that "roughly 25 per cent of normal children have occult defects in the vertebral laminae and this incomplete closure is frequently demonstrated in x-ray films taken for some other reason."

Sarpyener[9] has been a proponent of the relationship of congenital spinal canal stricture associated with spina bifida occulta in the development of a multiplicity of ortho-

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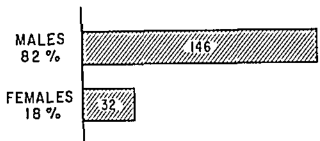



FIG. 1. Legg-Calvé-Perthes disease. Showing incidence in males and females.

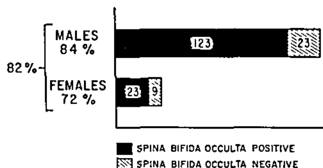



FIG. 2. Incidence of spina bifida occulta in Legg-Calvé-Perthes disease.

paedic conditions, including Legg-Calvé-Perthes disease and hip subluxations. He did not give statistics other than to show that spina bifida occulta coexisted with all the pertinent orthopaedic abnormalities.

Many references also exist in the literature relative to the incidence of spina bifida occulta in adults. These estimates vary from Southworth and Bersack,[10] who found 18.2 per cent in their series; Dittrich,[2] who found 5 per cent in his series; and Friedman, Fischer and Van Demark,[4] who, in their examination of normal soldiers, found 36 per cent incidence.

Brailsford,[1] commenting on defects of the neural arch, stated that its centers of ossification were present at birth and fused together during the first year. Epstein[3] agreed that "the laminae fuse at about the first year of life." Köhler[8] had an entirely different opinion, and the statement, "We may recall that the closure of the arch normally is terminated in males of the seventeenth year and in females in the fifteenth year," speaks for itself.

## PRESENT STUDY

The material for this report comes from 178 cases of Legg-Calvé-Perthes disease. These patients had previously received treatment or were still under treatment at the Blythedale Convalescent Orthopedic Hospital. Of these patients, 146 were male and 32 female (Fig. 1). Including all forms of spina bifida occulta, 123 of the 146 male patients showed some defect in closure of the neural arch at one or several levels in the lumbosacral region. Of the 32 female patients, 23 showed a similar lumbosacral laminal fusion failure (Fig. 2). The over-all incidence of spina bifida occulta in the entire group was 82 per cent.

The ages of the patients in this study ranged from 3 to 14 years, the average age being 6 years (Fig. 3). By breaking down

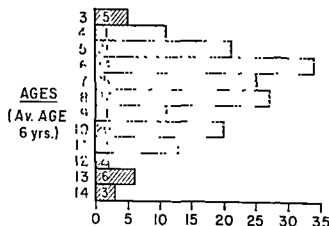



FIG. 3. Total number of patients with Legg-Calvé-Perthes disease.

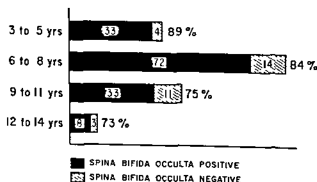



FIG. 4. Patients with spina bifida occulta in Legg-Calvé-Perthes disease by age group.

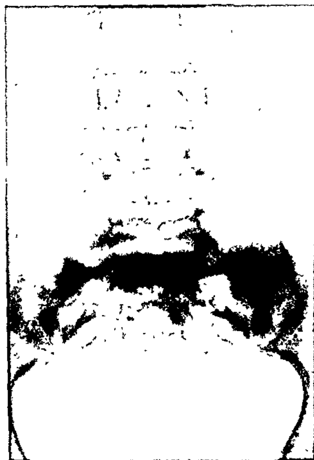


Fig. 5. S. M., female, 7½ years of age. Illustrates spina bifida occulta in Legg-Calvé-Perthes disease at S-1.

Table 1. Forms of Spina Bifida Occulta in Legg-Calvé-Perthes Disease

| | No. of Cases | Percentage |
|---|---|---|
| S1 .................... | 54 | 37 |
| S1-S2 ................. | 24 | 16 |
| L5-S1 ................. | 14 | 9 |
| L5 .................... | 12 | 8 |
| S1-S2-S3 .............. | 10 | 7 |
| L5-S1-S2 .............. | 6 | 4 |
| S1-S2-S3-S4-S5 ........ | 5 | 3 |
| Other assorted combinations .. .... .. | 21 | 16 |
| Total ...... ..... | 146 | |

the total number into separate age-group categories, it was found that the highest incidence of spina bifida occulta occurred in the 3- to 5-year age span, being 89 per cent, and the lowest in the 12- to 14-year age group, where it was 73 per cent (Fig. 4) This trend in diminution of the laminal defects with age corresponded to the findings of Hintze,[5] who was able to show it more convincingly, since his study included ages up to 17 years.

The patterns of neural arch defect in the patients with spina bifida occulta were multitudinous. The S-1 defect (Fig. 5) was that encountered most commonly, with the others listed in Table 1 in lesser incidence. This corresponded to the findings of previously published series[5,7] No relationship to Perthes severity existed

A second series of cases, also in residence at Blythedale, was analyzed. This category consisted of all varieties of orthopaedic disorders other than Legg-Calvé-Perthes disease. There was a total of 89 cases available for study. Sixty patients presented spina bifida occulta of various forms, representing a 67 per cent incidence. It should be mentioned that the mechanics of the study in this series differed from that which prevailed in the Legg-Calvé-Perthes group. In the latter, many roentgenograms were available for study. These proved to be of inestimable value, since no special preparation was used for optimum demonstration of the lumbosacral area. Gas shadows, poor technic and improper positioning were some of the obstacles encountered. With many roentgenograms at hand in Legg-Calvé-Perthes disease, the uncertainty of reading a single roentgenogram was eliminated. Figure 6 illustrated a case of scoliosis in which 2 roentgenograms were taken on the same date The one with the child in recumbency clearly shows spina bifida occulta of the first sacral segment, whereas the other, with the child in the sitting attitude, is obscured. The lower incidence of spina bifida occulta in this control group may be linked to the techni-

TABLE 2. FORMS OF SPINA BIFIDA OCCULTA IN MISCELLANEOUS ORTHOPAEDIC CASES OTHER THAN LEGG-CALVÉ-PERTHES DISEASE

| | No. of Cases | Percentage |
|---|---|---|
| S1 | 29 | 48 |
| S1-S2 | 10 | 16 |
| L5-S1 | 9 | 15 |
| L5 | 2 | 3 |
| S1-S2-S3 | 2 | 3 |
| L5-S1-S2 | 2 | 3 |
| S1-S2-S3-S4-S5 | 1 | 1.5 |
| Other assorted combinations | 5 | 10.5 |
| Total | 60 | |

cal detail of having only isolated roentgenograms for analysis.

The breakdown of forms of neural arch defect in this second miscellaneous orthopaedic group is listed in Table 2. The high incidence of S-1 defect is noted again. The similarity, in general, of the pattern of forms to those seen in Legg-Calvé-Perthes disease is obvious.

An unusual opportunity to trace the course of spina bifida occulta was available in 64 cases of Legg-Calvé-Perthes disease. The period of follow-up varied from 2 to 20 years after completion of treatment. In these 64 cases the initial incidence of spina bifida occulta was 81 per cent. On follow-up, the incidence of spina bifida occulta diminished to the level of 55 per cent. The average age of these Perthes children *initially* was 6.5 years; the youngest was 3 years; the oldest, 13 years. The average age at *follow-up* was 14.1 years; the youngest, 5 years; and the oldest, 27 years. The details of this group are shown in Table 3.



FIG 6. A. C., male, 5 years old, with paralytic scoliosis. (*Left*) Shows spina bifida occulta of S-1. This roentgenogram was taken with the child lying down. (*Right*) In this roentgenogram, which was taken immediately after the other, with the child in a sitting position, the defect is obscured because of the pelvic tilt.

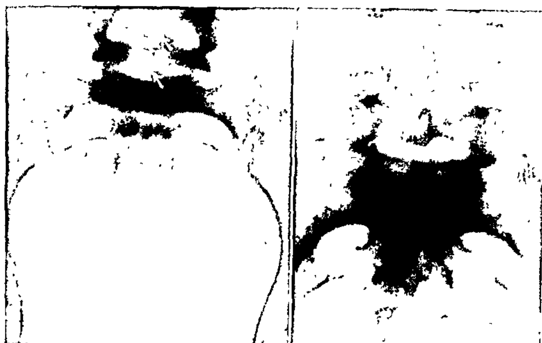


FIG. 7. T. E., female. (*Left*) Shows a faint cleft at L-5 and a defect at S-1 at 8½ years of age. (*Right*) Indicates closure at 13 years of age.

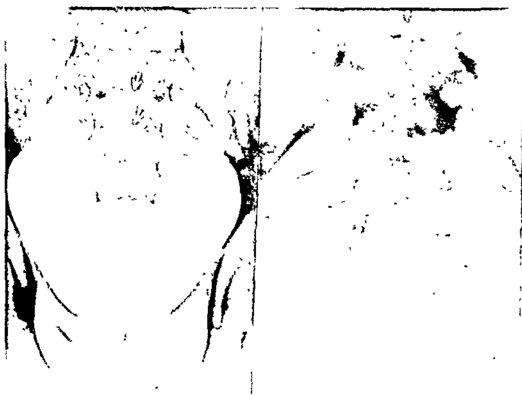


FIG 8 M J, male (*Left*) Shows spina bifida occulta with laminal defects of S-1, S-2 and S-3 at 6 years of age (*Right*) Indicates closure at 19 years of age.

FIG. 9 D. L., female (*Left*) Shows spina bifida occulta of S-1 at 7 years of age. (*Right*) Indicates closure at 24 years of age.

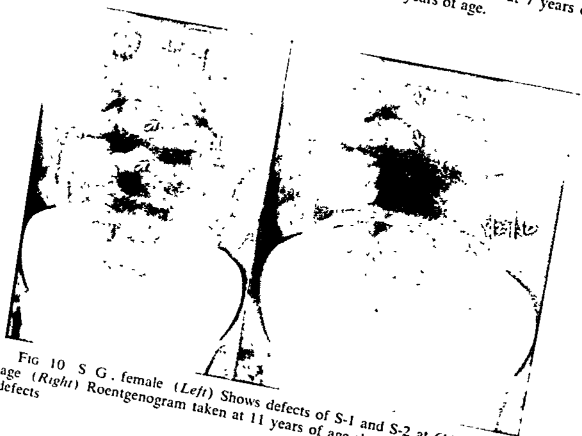


FIG 10 S G, female (*Left*) Shows defects of S-1 and S-2 at 6½ years of age (*Right*) Roentgenogram taken at 11 years of age shows persistence of the defects

TABLE 3. EXPERIENCE OF 64 CASES OF LEGG-CALVÉ-PERTHES DISEASE ON FOLLOW-UP

| | | |
|---|---|---|
| 10 | initially negative. | |
| | Neg. on follow-up .... | (av. age 7.2 years) |
| 2 | initially unknown. | |
| | Neg. on later follow-up | (av. age 20.5 years) |
| 35 | initially positive. | |
| | Pos. on follow-up .... | (av. age 13.8 years) |
| 6 | initially positive. | |
| | Neg. on short follow-up | (av. age 9.0 years) |
| 11 | initially positive. | |
| — | Neg. on later follow-up | (av. age 16.3 years) |
| 64 | | |

| | |
|---|---|
| Average age initially ............ | 6.5 years |
| Average age at follow-up ........ | 14.1 years |
| Incidence spina bifida occulta initially .. | 81% |
| Incidence spina bifida occulta on follow-up ...................... | 55% |

The major element producing the diminished incidence of spina bifida occulta was contributed by 17 patients. These cases showed laminal defects on initial examination but on re-examination indicated fusion of the neural arches. Six patients (Fig. 7) became negative on early follow-up at a time when their average age was 9 years. Eleven patients (Fig. 8) with initial failure of laminal fusion became negative after longer intervals, their average age at follow-up being 16.3 years.

The striking demonstration of spina bifida occulta at one age level, proceeding to closure upon further maturity, is noteworthy (Fig. 9). This observation had not previously been recorded, though its probable existence had been implied in the documentation of lower incidences of spina bifida occulta in older children and adults.

## DISCUSSION

The occurrence of spina bifida occulta (Figs. 10 & 11) continues to intrigue because of its inexplicability. In adulthood, when there is a relatively low incidence of this anomaly, it requires little imagination to accept it as a benign curiosity without serious significance. In children with high

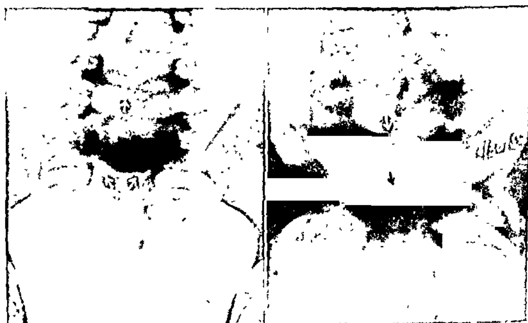


FIG. 11. E B, male. (*Left*) Shows spina bifida occulta involving L-5 and S-2, S-3, S-4 and S-5 at 6 years of age (*Right*) Roentgenogram taken at 23½ years of age shows persistence of the defects with prominent malformation of the spinous process of L-5

incidences of spina bifida occulta, there has been a greater reluctance to consider it a variation of the tempo of ossification. As a consequence, individual research workers have directed attention to possible linkage between spina bifida occulta and enuresis, as well as orthopaedic and neurologic abnormalities. The evidence of such association is wholly inconclusive when one considers that in normal children studies have shown similar incidences of spina bifida occulta. In Legg-Calvé-Perthes disease, an affliction confined solely to preadolescent children, the incidence of spina bifida occulta is high. There is a gradient diminution in this neural arch defect wth age, even in the childhood and the adolescent age spans, suggesting the orderly progression of maturity. In Legg-Calvé-Perthes disease, where follow-up is available, a greater diminution occurs as young adulthood is attained. It is extremely unlikely that the coexistence of Legg-Calvé-Perthes disease and spina bifida occulta have more than coincidental relationship.

## CONCLUSIONS

1. In 178 children with Legg-Calvé-Perthes disease between the ages of 3 and 14 years, the incidence of spina bifida occulta was found to be 82 per cent.

2. No correlation of spina bifida occulta was found to exist with the severity of the Perthes process.

3. Diminution in the incidence of spina bifida occulta occurred with advancing age as demonstrated by a follow-up study in 64 cases of Legg-Calvé-Perthes disease.

4. There is no conclusive evidence to show a definite relationship between spina bifida occulta and Legg-Calvé-Perthes disease.

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## Spina Bifide Occulte in Morbo de Legg-Calvé-Perthes

*Summario in Interlingua*


Esseva interprendite un studio de 178 casos de morbo de Legg-Calvé-Perthes pro determinar si o non il existe un association inter illo e spina bifide occulte. Le serie includeva 146 pueros e 32 pueras de etates de inter 3 e 14 annos. Le incidentia general de spina bifide occulte in le gruppo esseva 82 pro cento. Un grande varietate de defectos de arco neural esseva constatate. Localisationes a S-1 esseva le plus numerose. Le incidentia de tal defectos e le distribution de lor localisationes esseva simile a illos reportate in le litteratura pro juveniles enuretic e pro juveniles normal del mesme gruppos de etate.

Studios subsequente in 64 patientes de

morbo de Legg-Calvé-Perthes, effectuate inter duo e vinti annos post lor resanation, monstrava un reduction del incidentia de spina bifide occulte ab 81 usque a 55 pro cento. Es opinate que spina bifide occulte in juveniles representa un retardo del ossification sin signification pathologic. Un diminution considerabile del condition occurreva naturalmente in le curso del maturation usque al stato adulte.

## Section II

# GENERAL ORTHOPAEDICS



# 13

# Surgical Endeavors in Arthritis

H. KELIKIAN, M.D., S. SARAFIAN, M.D., L. TOPOUZIAN, M.D., AND HRATCH DOUMANIAN, M.D.*

Diseased and disabled joints present a variety of problems, some of which fall within the domain of surgery. The orthopaedic surgeon often is asked to confirm a diagnosis—aspirate and inject a joint and perform a biopsy—and he has at his disposal a number of surgical procedures which, with judicious application, will give the arthritic patient a measure of relief from pain, correct his deformity, or enhance the movements of his joint or stabilize it.

## DIAGNOSIS

It is not uncommon to find a joint that has been dubbed arthritic and treated as such for years to be the seat of some other affliction—tumor, for instance. Moreover, there exists considerable confusion as to how the two main varieties of nonspecific arthritides—rheumatoid and osteoarthritis—manifest themselves and in what respect they resemble or differ from tuberculous or pyogenic involvement and from trophic joint diseases.[1,4,5]

**Rheumatoid arthritis** is a disease of the relatively young, usually it starts before 40. It is more common in women and in people of northern European extraction. It is a generalized disease: at one time or another it will manifest such systemic signs as elevated temperature, leukocytosis and increased sedimentation rate. It may be accompanied by myocardial or valvular heart disease and subcutaneous nodules, and usually it affects more than one articulation. It has a predilection for joints with extensive synovial lining and bulbous articular ends—joints rich in vascular connective tissues as are found under the synovial surface and in the subchondral marrow spaces. Taking the hand as an example, rheumatoid arthritis chooses the metacarpophalangeal and the larger proximal interphalangeal joints. Rheumatoid arthritis leads to marked deformity, dislocations and, at times, ankylosis. Roentgenograms will show the bones to be porotic and the cartilage space between them attenuated and, not infrequently, effaced. At arthrotomy, one finds the synovial membrane turgid, congested, corrugated, thrown into folds and rugi, warted with multiple villi or bound down with adhesions; the articular cartilages are eroded from the periphery, where the synovial membrane overlaps the margin of the hyaline cartilage—the inflamed synovial membrane creeps over and eats away the more specialized connective tissue, i.e., the articular cartilage. This is a situation in which relatively primitive, vascular, hence *inflammable,* connective tissue displaces the specialized elements of the joint, i.e., hyaline cartilage and bony trabeculae.

**Osteoarthritis** is primarily degeneration of specialized elements of the joint, the articular cartilage in particular. It is a localized affliction. Except in the spine, usually

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it affects a single, solitary joint—at most two, rarely three. It is a wear-and-tear process. It occurs most commonly in robust laboring adults, in the old and the obese, in connection with deformities, misalignment and muscular imbalance, and following trauma. Osteoarthritis has a predilection for weight-bearing overused articulations and for joints far away from the central circulation, hence poor in it, as, for example, the terminal interphalangeal articulations of the hand. Senility is not only protracted trauma, it is also a drying-up process; with it there is arteriosclerosis and progressive diminution of the blood supply of the joints, especially the terminal joints of the hands and the feet. The osteoarthritic joint may be limited in movement, but this is due to mechanical incongruity or interlocking, not to frank ankylosis. Roentgenograms may show widening and mushrooming of the articular ends of bones, condensation or subchondral cortex, cystic vacuolization of subjacent metaphysis, osteophytes and osteocartilaginous loose bodies. At arthrotomy, usually one notes ulceration of the central portion of the hyaline surface and spur formation of the margins; the synovial membrane is uninflamed except at the chondrosynovial junction, where it may appear to be turgid due to frictional irritation by abutting spurs.

**Tuberculous arthritis** resembles rheumatoid arthritis in that it may be accompanied by systemic signs. Then, too, the destruction of the articular surfaces is most extensive around the periphery or at points of noncontact and pressure where weaker tuberculous granulations coming from synovial membrane can survive; the articular surfaces are eroded by these marginal granulations and by inflammatory process in subchondral marrow spaces. In contrast with rheumatoid arthritis, tuberculosis rarely involves more than one joint, at most two, the roentgenograms may suggest calcification of tuberculous exudate and sequestrations of portions of articular ends of bones, so-called kissing sequestrae, neither of which occurs *in rheumatoid arthritis* Tuberculous exudate breaks through and forms extracapsular abscess and sinus. It causes no tissue reaction in the form of induration, surface heat (hence the name *cold abscess*) or redness. It has practically no proteolytic action. Tuberculous pus contains monocytes and is alkaline in reaction. What little enzyme monocytes produce is inactive in alkaline medium; therefore, tuberculous pus has no digestive power. Destruction in tuberculous arthritis is caused mainly by granulations; it is gradual, slow. Diagnosis is confirmed by guinea-pig inoculations and tissue biopsy.

**Pyogenic arthritis** simulates osteoarthritis in that the greatest erosion of articular surfaces is at points of contact and pressure, and the ulceration of the hyaline surface is located more centrally. But, of course, pyogenic arthritis has an acute onset and is accompanied by fever and leukocytosis Eventually it leads to ankylosis, which is not the case in osteoarthritis. *In contrast* with tuberculous pus, pyogenic exudate i acid in reaction; it is rich in leukocyte which produce a proteolytic enzyme that i active in acid medium. In pyogenic arthriti the articular surfaces are destroyed rapidl by the digestive action of leukocytic en zymes. Pyogenic pus also breaks out int the surrounding tissues, but, unlike tuber culous exudate, it causes local reaction i the form of induration, surface heat and red ness. Sometimes there is even bacteremia as confirmed by blood cultures.

**Pigmented villinodular synovitis** ma simulate the villous phase of rheumatoi arthritis. It may throw an apron over th articular cartilage, but it causes no erosion the hyaline surfaces under the synovial expansion remain intact. The villi may bruis one another, necrotize and cause intermittent bleeding, lending a pinkish hue to the joint fluid. Reabsorbed blood imparts to the synovial membrane the peculiar pigmented appearance. In pigmented villinodular synovitis the aspirated joint fluid is brown, serosanguineous or bloody. After the introduction of air into the joint, arthrograms[7] will *show a smooth unbroken contour* of the

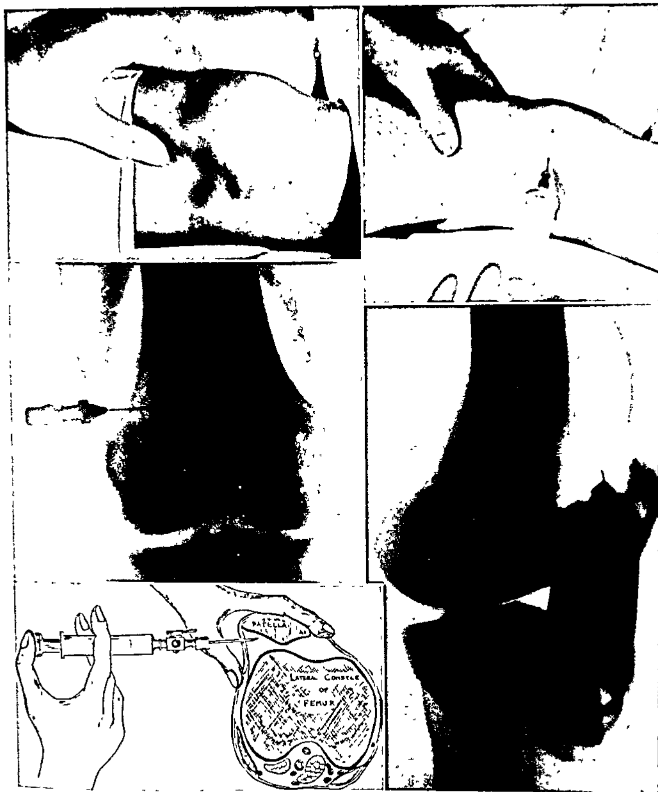



FIG 1. Aspiration and injection Insertion of the needle into the articular cavity serves several purposes: excessive fluid is aspirated, thereby relieving the intra-articular tension; aspirated fluid is subjected to chemical and cytologic study; air is introduced into the joint and arthrograms are taken; finally, Hydrocortone is instilled into the articular cavity.

(*Top, left*) Showing the displacement of the patella laterally and insertion of the needle into the knee through the tautened lateral patellofemoral ligament or capsule. (*Top, right*) The same with the syringe connected. (*Left, center*) Anteroposterior roentgenogram showing the needle under the shadow cast by the patella. (*Bottom, ri* Arthrogram: introduction of air into the articular cavity. (*Bottom, left*) the displacement of the patella laterally and the insertion of the needle.

joint cavity, whereas in rheumatoid arthritis the shadow cast by the joint outline may be ragged. The same is true in all inflammatory arthritides, and this irregular outline denotes adhesions, the aftermath of inflammation.

**Synovial chondromatosis**[6] and **osteochondromatosis** are somewhat akin to pigmented villinodular synovitis. They probably are not caused by inflammation and are due to metaplasia of the primitive connective tissue elements. Cartilaginous or osteocartilaginous plaques are laid within synovial folds or villi, hanging like bunches of grapes or breaking loose and floating in the joint cavity. Detached pieces may cause frictional erosion of the articular surfaces and create a condition not unlike osteoarthritis. Roentgenograms, with or without introduction of air, will show the true state of affairs: osteocartilaginous bodies can easily be seen on roentgenograms, and hyaline plaques will be recognized as space-occupying translucencies or filling defects in arthrograms.

There are other joint disturbances to be differentiated, such as gouty arthritis, hemophilic joint, diabetic and tabetic arthropathies, gonorrheal arthritis, allergic and syphilitic arthritides. Usually, the diagnosis of any of these conditions is arrived at by associated clinical findings, roentgenographic studies and laboratory tests. Definite diagnosis of joint tumors can be made only by biopsy.

## SURGICAL MEASURES

If operative interference in joint disease has accomplished nothing else, it has brought to light numerous conditions that have very little or nothing to do with arthritis. By surgical interference we mean arthrotomy and biopsy. In many instances a simpler measure, such as joint aspiration, will furnish a clue. Then, too, with a needle in the joint, air may be injected and arthrograms taken that may give additional information as to the contour, the capacity, the content and the communications of the articular cavity.

**Joint aspiration** constitutes the simplest surgical approach toward solution of a given problem in connection with arthritis. Not only does it relieve pain by reducing the tension caused by excessive accumulation of fluid, but, by the characteristics of the fluid obtained, such as reaction, smears, culture, cell count, uric acid content and the effect on the inoculated laboratory animal, it gives a clue as to the exact nature of the disease affecting the joint. At times joint aspiration is followed by injection of Hydrocortone and, when infection is suspected, by the instillation of a suitable antibiotic (Fig. 1).

**Arthotomy** is done for purposes of biopsy, for the removal of osteocartilaginous loose bodies, for the eradication of articular incongruities and for the smoothing out of spurs, and it goes without saying that it is the first step in such major procedures as synovectomy, arthroplasty arthrodeses, resection of articular ends, ostectomy or sesamoidectomy, such as excision of the patella in cases of advanced osteoarthritis. Reduced to its true connotation, arthrotomy is the making of a small aperture in the joint for purposes of biopsy, for evacuating the products of inflammation or degeneration, or for extracting an osteocartilaginous loose body which, left alone, might cause frictional irritation of the articular surfaces and pave the way to osteoarthritis. Not infrequently the process pursues a reverse course; in osteoarthritis, osteophytes may become detached and float in the joint. There are instances in which the osteocartilaginous body within the articular cavity is definitely the result of synovial metaplasia. Itself originating from mesoderm and remaining more or less primitive, the synovial membrane can, by metaplasia, produce the more specialized end-products of connective tissue; i.e., cartilage and bone. This tendency is at times familial; one often sees osteocartilaginous loose bodies in the knees of various members of the same family, which is also true in pigmented villinodular synovitis.[5,7] Osteocartilaginous loose




Fig 2. Arthrotomy In a broader sense arthrotomy is the first step in most major surgical attacks on the joint proper. Reduced to its more confined connotation, arthrotomy denotes making a small aperture in the joint and removing a piece of tissue for purposes of biopsy or extracting the by-products of inflammation, injury or metaplasia

(*Top, left & right*) Anteroposterior roentgenograms of the right and the left knees of the same individual with bilateral osteocartilaginous loose bodies (*Bottom, left*) Arthrogram of the knee shown at *top, right.* (*Right, center*) Arthrotomy and extrusion of an osteocartilaginous body from one knee (*Bottom, right*) Osteocartilaginous bodies extracted from both knees. The scale is in centimeters

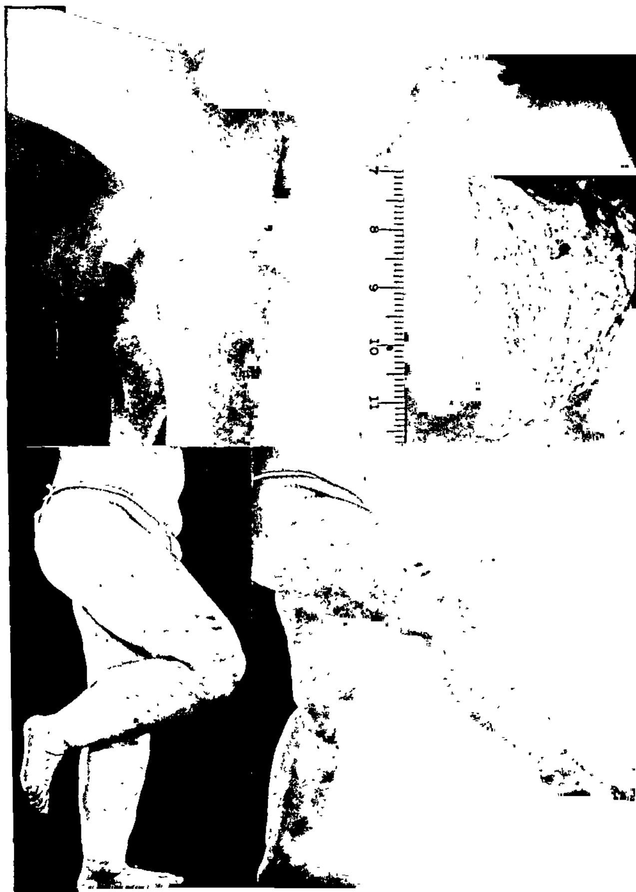


FIG 3. (*Caption on facing page*)

bodies may occur bilaterally in both knees of the same individual (Fig. 2).

Osteotomy is performed to correct deformities caused by arthritis, to shift the line of stress and thereby alleviate pain, and to enhance the functional usefulness of the segments connected at the joint. Osteotomy may accomplish the desired result by dis-

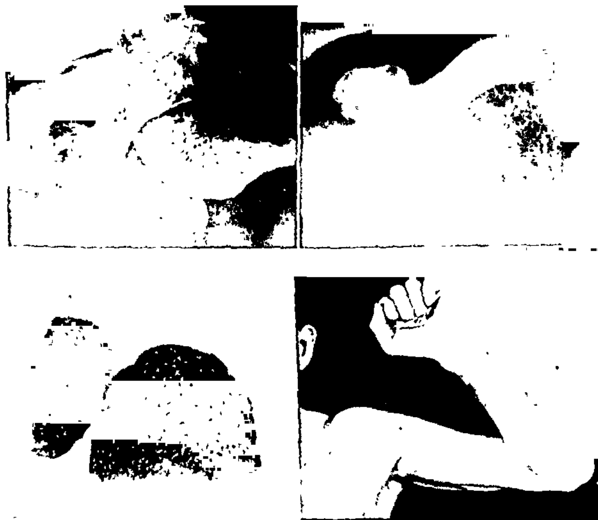


FIG. 4. Resection of the articular ends of both bones entering into the formation of a joint. (*Top, left*) Roentgenogram of the acromioclavicular joint before resection. Note the arthritic change. (*Top, right*) The same after resection. (*Bottom, left*) Roentgenogram of the resected portions of the clavicle and of the acromial process of the scapula. (*Bottom, right*) Showing the range of postoperative painless abduction.

FIG. 3. Total ostectomy or sesamoidectomy Excision of the patella is now a standard procedure for chondromalacia or osteoarthritis of this largest sesamoid bone. The condition is most common in knock-kneed obese women: as the knee is flexed the patella rides over the lateral condyle of the femur and wears itself out by unwonted friction—not gliding in its usual groove.

(*Top, left*) Lateral view roentgenogram of the knee of a woman in her sixties. (*Top, right*) Transpatellar or "sky view" of the same. Note that the patella overrides the lateral femoral condyle (*Center, right*) The resected patella. (*Bottom, left & right*) Functional results.

placing, angulating and rotating one of the surgically severed fragments of bone. In some instances several maneuvers have to be combined: rotation with displacement and the latter with angulation. Rarely is osteotomy practiced on the spine; occasionally for deformities of the elbow, the wrist or the fingers.[3] Osteotomy finds wider application for such weight-bearing joints as the ankle and the knee, and especially for the hip. In osteoarthritis of the hip, subtrochanteric osteotomy[7] of the femur attempts to accomplish several purposes: by angulation or displacement of the distal fragment it proposes to change the line of stress; by controlled rotation of the proximal bone it aims to bring a comparatively unworn articular portion of the head of the femur under the weight-bearing sector of the acetabulum.

*Ostectomy* may be partial or complete. An example of total ostectomy is the removal of one of the sesamoid bones; the patella, for instance. In chondromalacia of this largest sesamoid bone, one may hesitate between remodeling it and removing it *in toto*. In the young, it is perhaps wiser to reduce the size of the patella, leaving an osseous core that will maintain the continuity of the quadriceps pull. This is especially true in cases of chondromalacia of the patella due to recurrent dislocation. At the same sitting that the insertion of the patellar tendon is shifted medially on the proximal tibia the patella is reduced in size, shaved along its medial and lateral borders, and remodeled on its articular aspect, leaving unmolested the superior and the inferior poles where the tendons connect. Then the denuded articular surface of the patella is covered with folds of capsular tissue much after the manner of peritonealizing a viscus. In the aged, in whom degenerative arthritis is more common, the patella often is mushroomed and interlocked, and one must forego such refined procedures as patellaplasty and resect the patella as a whole (Fig. 3).

Partial ostectomy or resection of
ends is also practiced in non-weigh
joints: in the jaw, the condyle and t
noid process of the mandible are
in the acromioclavicular joint, the o
of the clavicle; in the elbow, the he
radius; in the distal radio-ulnar arti
the terminal inch of the ulna; in t
one or all of the carpal bones of t
mal row; in the hand, the heads of t
carpals; in the fingers, the distal en
proximal phalanges. In arthritic fe
an inch or more of the distal end
middle 3 metatarsals are resecte
preferable not to remove the end
first and the fifth metatarsal bones
stead, to resect the proximal portio
adjacent phalanges. Phalangectomy
or complete, is practiced at times
formities of the toes: phalangectom
4 lateral toes leaves them flail, an
this way, they will shrivel backw
become unsightly, even painful. It
able surgically to syndactylize each
adjacent toes together.

Partial ostectomy finds its valid
tion in mobilization of the temporom
lar joint and in painful arthritis at t
mioclavicular junction, both pure
weight-bearing articulations (Figs.

*Synovectomy* may also be partial
plete. The best example of partial s
tomy is in the resection of the gastroc
semimembranosus bursa, or Bake
Both these terms—*cyst* or *bursa*—
misapplied. What we actually hav
outpouching, or diverticulum, of t
joint that may be pathologically e
the sac on the medial aspect of the p
space may swell as a part of the gen
change, affecting the lining of the
articular cavity of the knee; or, bec
its dependent position, which favors
or stagnation, it may be the discrete
disease. In the latter instance, extirp
the popliteal sac will suffice; but, w
swelling is a part of generalized dis
the joint lining, synovectomy has to



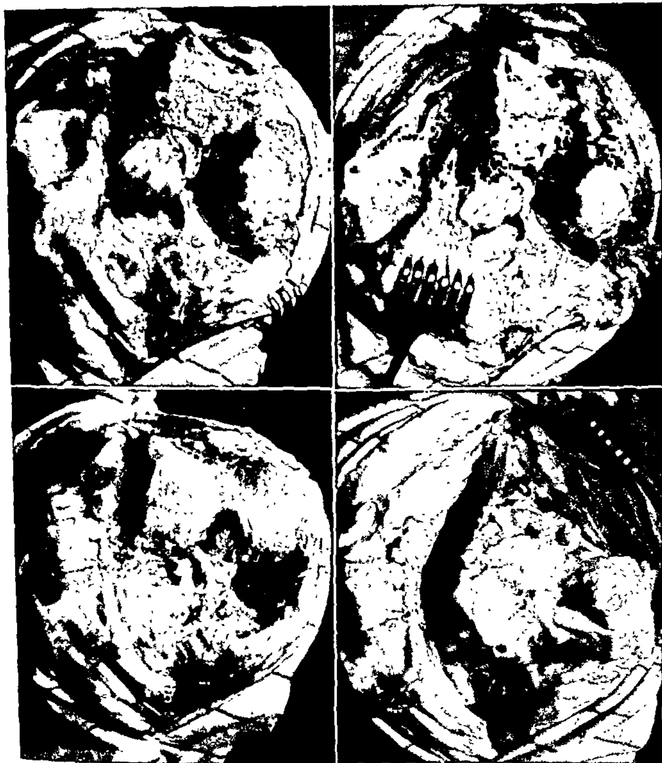
Fig 5. Mobilization of a joint by resecting one of the articular ends forming a joint. (For details of this operation see Ref. 2, p. 138.) (*Top, left*) Frontal view of a woman with interlocked temporomandibular joint. (*Top, right*) Profile view of the same (*Center, left & right*) Showing the range of mobility of the lower jaw before surgery. The scales are in inches. (*Bottom, left & right*) The same after resection of the articular ends of the mandible. The pimplelike discoloration on the chin seen at *right* is due to the Steinmann pin used for traction postoperatively.



FIG 6. Arthrolysis for fibrous ankylosis combines the principles of synovectomy with those of dissection of scars and bands In the knee, scar-buried menisci, and even the cruciate ligaments, are removed; in the suprapatellar pouch the dissection is carried down to the bare cortex of the distal articular end of the femur

(*Top, left*) The knee of a woman with complete fibrous ankylosis due to supracondylar fracture. Preoperatively there was only 10° passive painful motion. The probe passes a thick band of scar binding the patella to the femoral condyle. (*Top, center*) The same after release of the patella showing the scar-matted articular surface and the scar-buried cruciate ligaments. (*Top, right*) The same after completion of dehiscence Note that even the periosteum has been stripped, and nutrient foramina are evident (*Bottom, left, center & right*) Functional results. This patient has been followed now for 8 years The knee is painless and stable: "cruciate ligaments are not crucial"

complete. Perhaps *complete* is too strong a word; subtotal is nearer the truth. When synovectomy is contemplated for old burnt-out rheumatoid disease, it is best to ascertain first the state of inflammation in the joint In the knee, a few cubic centimeters of oil is introduced into the joint, and lateral view roentgenograms are taken at intervals. If, within a month, the popliteal lymph node becomes impregnated with oil and is visual-



FIGS. 7 and 8 Arthroplasty, or adaptive remodeling of the articular ends of bones, finds its greatest usefulness in the elbow and the hip. In the knee it is to be regarded as a compromise procedure—not nearly as beneficial as arthrodesis.

FIG. 7. (*Top, left & right*) Show somewhat different views of the knee of a woman with burnt-out rheumatoid arthritis Note the destruction of the articular surfaces of the femoral condyles. (*Bottom, left*) The same after remodeling. (*Bottom, right*) The filed-down femoral condyle has been covered with a pedunculated fascial flap obtained from around

ized in roentgenograms, it means that the synovial lining readily absorbs the radiopaque substance, hence is hyperemic—hot—and the joint had better not undergo operation.[1,4,5] We depend more on this test than the sedimentation rate, which may be increased due to inflammation elsewhere or in some other joint than the one under consideration.

**Arthrolysis**, or dehiscence of scar-bound articular surfaces, is somewhat akin to synovectomy: not only are adhesions severed but also scar-producing synovial lining is dissected out. In the more severe cases of fibrous ankylosis one must do more; one must extirpate the intra-articular fibrocartilage as the menisci of the knee. In the suprapatellar pouch of this joint, where most of the adhesions occur, the dissection is carried to the undersurface of the quadriceps muscles on the one hand and on the other down and including the periosteum surrounding the distal articular end of the femur (Fig. 6).

**Arthroplasty**, or adaptive remodeling of opposed articular surfaces, is an operation that finds its greatest usefulness in the fingers, the elbow and the hip. The primary purpose is to enhance movement, which is of paramount importance for the upper extremity. Preliminary to mobilization of the joint, one must ascertain the power of the muscles moving the segments connected at the joint—make sure that they have not wasted away by prolonged lack of use or turned fibrotic In weight-bearing joints, arthroplasty is used most often for the hip. At this joint, where a globular head has to be carved from one flattened and mushroomed, and sometimes the acetabulum has to be deepened, the lever between the attachment of muscles to the greater trochanter and the axis of the joint movement is diminished; the trochanter rides high, and the tension of the attached muscles is nullified. Muscular function can be improved greatly by shifting the trochanter downward on the shaft, by combined angulation-type subtrochanteric osteotomy or by an endoprosthesis of conferrable head and neck to procure the desired leverage.[5,8]

Arthroplasty denotes a reshaping of the opposed articular ends of bones and interposition of some such material as will prevent reunion of the freshened surface. The knee is not a ball-and-socket joint, as is the hip, that lends itself to adaptive remodeling; nor is it as deeply situated or swathed with heavy musculature and able to tolerate the interment of foreign material in the form of plates, caps or prostheses. In the knee, arthroplasty is less likely to succeed, and arthrodesis is the better operation. Occasionally, however, arthroplasty of the knee is attempted as a compromise measure. In a woman of sedentary habits with no economic exigency, with a stiff hip on the same side making such a simple performance as slipping on a stocking an impossible feat, one is justified in mobilizing the knee, hoping that this will permit some little motion and remain relatively painless. No surgeon in his right mind should perform arthroplasty on a knee that has undergone solid bony ankylosis and is in good functional position for walking. Arthroplasty is tried only on a knee that has become interlocked because of irregular destruction of articular surfaces; the incongruous grooves and gutters are smoothed out and covered by a pedicled fascial flap obtained from the vicinity (Figs. 7 & 8).

**Arthrodesis** is the most useful operation for such weight-bearing joints as the hip, the knee and the ankle. In unilateral hip involvement in young laboring men who earn their livelihood in occupations that require

FIG. 8 (*Top, left*) Lateral view roentgenogram of another woman with burnt-out rheumatoid arthritis, interlocked knee She had bony ankylosis of hip on the same side. (*Top, right*) The same after remodeling the femoral condyles and the patella. (*Center, left & right*) The knee during exposure (*Bottom, left*) Postoperative range of flexion (*Bottom, right*) Postoperative range of extension.

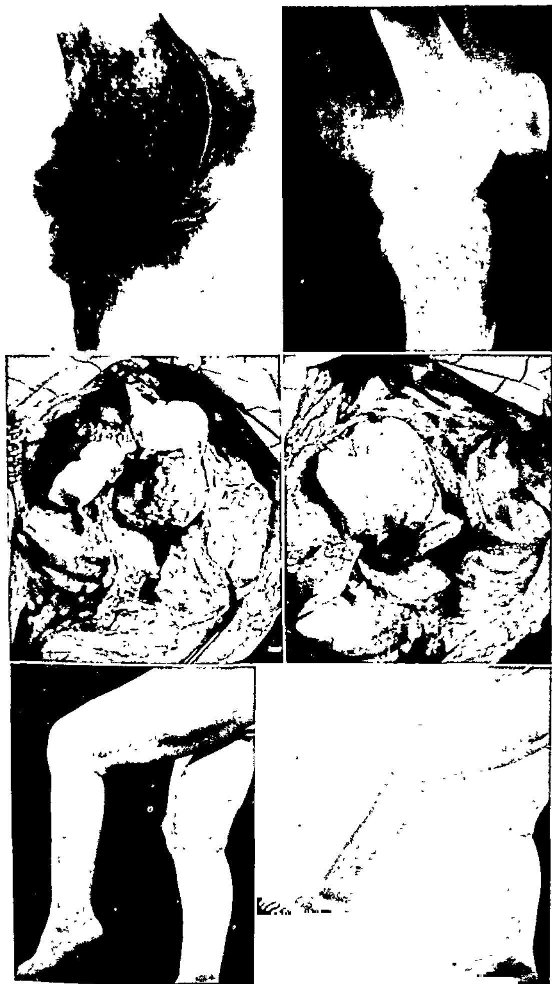


FIG. 8. (*Caption on facing page*)

standing or walking, it is the operation of choice. Occasionally, the interphalangeal joints of the toes are fused for painful hammertoes. In the upper extremity, the shoulders, the elbow, the wrist and the metacarpophalangeal joint of the thumb and, occasionally, the interphalangeal joints of the small finger may have to be stabilized.

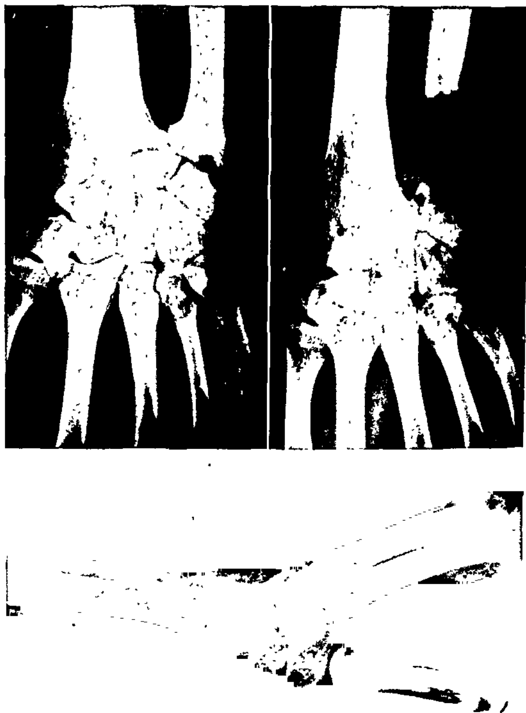


FIG 9. Arthrodesis. For weight-bearing joints no operation gives as gratifying results as fusion. In the wrist, too, it is to be preferred to carpectomy, especially in arthritis. (*Top, left*) Anteroposterior roentgenogram of the wrist of a woman with rheumatoid arthritis. (*Top, right*) The same after transulnar fusion the segment of resected ulna was used as a dowel graft between radius and os magnum (*Bottom*) Lateral view of the same showing the position at which the wrist is fused.

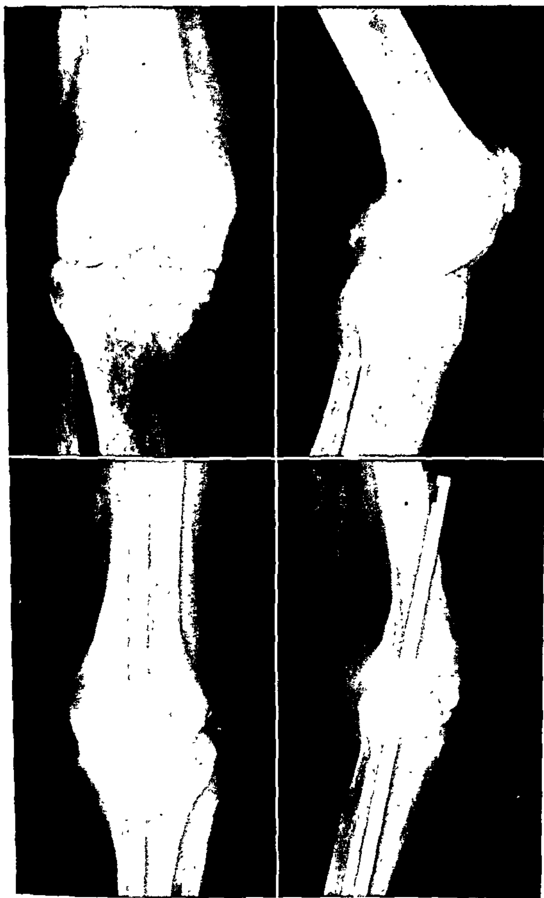


Fig. 10. Fusion of arthritic knee with the aid of an intramedullary rod. (*Top, left & right*) Preoperative anteroposterior roentgenogram of the knee of a woman with painful rheumatoid arthritis (*Bottom, left & right*) The same 6 months after surgical fusion and insertion of intramedullary rod.



FIG. 11. Transfibular arthrodesis of the ankle. (*Top, left*) Showing the exposure of the distal 3 inches of the fibula through a lateral incision. (*Top, center*) Passing the bone retractors under the shaft of distal fibula. (*Top, right*) The fibula has been resected, exposing the ankle joint (*Bottom, left & center*) Denudation of the opposed articular surfaces of the tibia and the talus and preparation of a groove into which the decorticated portion of the resected fibula is countersunk. Sometimes, through a small counter incision beneath the medial mallus, arthrodesis is completed. (*Bottom, right*) Closure.

When the elbow is ankylosed surgically, the proximal end of the radius is resected; when the wrist is fused, an inch or two of the distal ulna is resected to preserve pronation and supination of the forearm (Figs. 9-12).

## SUMMARY

Surgical measures most useful in arthritis are aspiration and injection of joints, arthrotomy, ostectomy, osteotomy, synovectomy, arthrolysis, arthroplasty and arthrodesis.



Fig. 12. Arthrodesis of the ankle. (*Top, left*) Anteroposterior roentgenogram of the ankle of a woman with painful arthritis. (*Top, right*) Lateral view of the same. (*Bottom, left*) Anteroposterior roentgenogram of the same 6 months after transfibular fusion. (*Bottom, right*) Lateral view of the same.

Not the least of the benefits derived from surgical interference in arthritis is the establishment of diagnosis and discovery of conditions other than arthritis, especially tumors.

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## Mesuras Chirurgic in le Tractamento de Arthritis

*Summario in Interlingua*


Varie mesuras chirurgic pro le tractamento de arthritis chronic es describite. Istos include aspiration e injection, arthrotomia, osteotomia, ostectomia, synovectomia, dehiscentia o elimination de adhesiones, ablation de osteophytos e incongruitates mechanic, e arthroplastia e arthrodesis.

# 14

# Pathogenesis of Lumbar Disk Lesions

L. STANLEY SELL, M.D.*

To any physician practicing in an area where farming, ranching, mining, lumbering or other heavy manual labor occupies most of the male force, it must sooner or later become evident that there is a correlation between type of work and lumbar disk injuries.

In reviewing the 4,000 records which comprise the background for this report, I was impressed by the histories of long-existing back difficulty, even in young adults. Friberg† states:

> Disk degeneration is very common, often appearing at an early age, but is not detected clinically until it has reached an advanced stage.

I believe that early post-traumatic disk degeneration can be detected on a basis of suspicion by the history of the injury and the symptoms, this is the time for long consultation between doctor and patient, for frequently the future of the patient's back depends upon what is said and done at this moment. There are scores of excellent articles and learned discussions on the anatomy, the physiology, the diagnosis and the treatment of lumbar disk lesions, but very little has been written that even hints at the prevention of these same lesions. Surely our knowledge has progressed enough in the past 22 years for us to use some of it in the prophylaxis of this condition. None of us is so egotistic as to believe that his treatment of an injured lumbar disk would be superior to the prevention of the injury in the first place.

A consideration of the etiologic factors in proven cases of lumbar disk herniation may prove useful in delineating what I have chosen to call the "natural history" of this condition.

Among high-school boys feeling the first proddings of manhood, weight-lifting contests have appealed as a proof of physical superiority. J. Albert Key reported an operative case where a weight-lifting contest was the etiologic agent in a ruptured lumbar disk, and I have had two of my own. Acrobatics on a trampolin are taught in some schools as a part of the gymnastics class, and here the acute hyperextensions have wrought not a few disk injuries. In like manner, back dives and flip-overs in the swimming pool have brought teen-agers to grief with their backs. Too early exposure to heavy work, such as "bucking spuds" in our area where teen-age youngsters work in harvesting the potato crop, has produced the first insult to a lumbar disk. Obviously, this type of exposure to injury is regional in occurrence, but other types of exposure may come to mind to those from other parts of the country where growing youngsters are exposed more or less deliberately to too heavy manual tasks If we physicians know that the foregoing are harmful to our children, would not some well-directed advice be as useful and as justifiable as the giving of antipolio vaccine?

* Idaho Falls, Idaho

† Friberg, S. Lumbar disc degeneration in problem of lumbago sciatica (Sir Robert Jones lecture), Bull Hosp. Joint Dis. 15:1-20, 1954.

Etiologic agents in adults are many and varied, but let us consider again some of those which have yielded proven cases of herniation and rupture of a lumbar disk. In car accidents, occupants may be thrown under the dashboard or catapulted from the front to the rear seat with acute herniation of a lumbar disk as the result. Prophylaxis here is too simple for adoption; namely, the seat or lap safety belt. I could devote the rest of this chapter to this favorite subject of mine, but it has already been developed admirably in other numbers of *Clinical Orthopaedics*. That we physicians have failed to educate our patients in this important safety measure is revealed by a statement by the Ford Motor Company that only 4 per cent of new cars ordered from the factory are accompanied by requests that safety belts be installed. Men lifting heavy truck tires in the act of changing them without any mechanical aid have damaged lumbar disks. Lifting trailer tongues to the coupling without the aid of a jack has proven hazardous. Stacking 90-pound bales of hay or loading 100-pound sacks of potatoes into a freight car using the back as a human derrick has damaged a frightening number of lumbar disks. Women lifting couches or beds unaided have come to grief. Nurses lifting heavy patients do more than "strain" their back muscles Anyone pushing a stalled automobile or trying to lift apart bumpers that are hooked is courting a serious injury to one of his lumbar disks. Lifting heavy overhead garage doors that are recalcitrant is a problem that would be better solved by driving through the door and replacing it with one that works easier!

Sir Arthur Keith,* writing on man's posture, states.

In none of the orthograde forms is such a continuous and urgent demand made on the postural spinal mechanisms as in man, with the whole weight of the supra-sacral portion of the body supported erect on the spine over long intervals. In fatigue the muscles which act on the short levers of the spine yield first, while the muscles which act on the long costal levels still keep on.

Thus the lumbar segments and their disks get the first results of the fatigue pattern, and the obvious medical implication here is to make some effort at changing those working conditions which fatigue the lumbar musculature. Keith concludes:

It is *not* true to say our spines are not perfectly adapted to the upright posture; it would be more accurate to say that human spines were not evolved to withstand the monotonous and trying postures entailed by many modern jobs and industries.

Goff,† in his excellent contribution "Orthograms of Posture," demonstrates how the lumbar lordosis of the muscular body type is the greatest, and so is the incidence of backache and degenerative disks in this type than in any other, with degeneration following the use of the part beyond its mean tolerance. Thus the muscular Tarzan thinks that he has to justify his size by performing muscular feats, and often the little man tries to keep up in order not to be outdone. All too frequently both end up with damaged lumbar disks.

Snedecor,‡ in his clinical study of industrial back injuries, found that under the mechanisms of injury, overstrain from fatigue produced a particularly bad group, with 50 per cent of these cases becoming chronic. These chronic cases became the problem cases; i.e., it was more difficult to treat them, much time was lost from work, and a relatively high percentage had permanent disability. A high percentage of recurrences of strains showed positive roentgenograms such as unstable lumbosacral joints due to disk degeneration, osteoarthritis and spondylolisthesis. If all these findings are

* Ellis, J O (Ed ) The Injured Back and Its Treatment, A Symposium, Springfield, Ill., Thomas, 1940.

1955.

true, could not pre-employment roentgenograms be used early to place the man properly in industry with a view to his own ultimate good? For those among us who act either as industrial physicians or as consultants should stimulate the development of mechanical devices to aid the human back in industry. We may also advise on changing working conditions harmful to employees' backs.

Remember that, to a high degree, symptoms are signs of a local aging, and if you cannot change the man, maybe you can alter his working conditions. Here is a vast field for rehabilitation in a broad sense—to find out what the man is capable of doing, to create better contact between doctors and employers, and to stimulate inventions in order to ease jobs, be they in the plant, the office or the home. Rehabilitation is a medical as well as a social responsibility and pays for itself. The individual goes off public assistance, returns to work and becomes self-sustaining; he may even be productive enough to pay taxes.

Doctors of medicine had a chance to see early and to diagnose properly 67 per cent of patients with back complaints (Table 1).

TABLE 1. EARLY MEDICAL CARE

| PERSONS CONSULTED | PERCENTAGE OF CASES SEEN IN EARLY STAGES |
|---|---|
| Physician | 45 4% |
| Chiropractor and osteopath | 32 4% |
| Physician; chiropractor and osteopath | 22.0% |

TABLE 2. DIAGNOSIS OFFERED PATIENTS BY PHYSICIANS IN ORDER OF FREQUENCY

1. Sacro-iliac conditions (slipping, subluxation or sprain)
2. Arthritis, neuritis, myositis
3. Infection (teeth, tonsils, prostate)
4. Endocrine dysfunction (thyroid, uterine, ovarian)

TABLE 3. TYPES OF TREATMENT GIVEN EARLY DISK CASES BY DOCTORS

1. Sacro-iliac belts
2. Back strappings
3. Massage
4. Procaine, local injection
5. Procaine, intravenous injection
6. Salicylates
7. Gold, *intramuscular injection*
8. Cortisone
9. Vitamin $B_{12}$
10. Heat lamp
11. Diathermy
12. Built-up shoe
13. Deep x-ray therapy
14. Prostatic massage
15. Antibiotics
16. Sacro-iliac fusion
17. Removal of: *Tonsils*, Teeth, Uterus, Ovaries

(The poor results obtained necessitated eventual surgical intervention.)

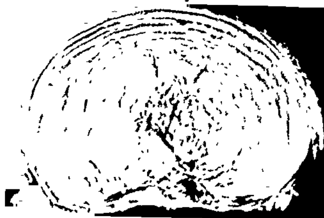


FIG. 1. Photograph of a ruptured disk from a patient with disk symptoms whose roentgenograms were negative. Note the central fracture with formation of almost a sequestrum in the posterior portion. (Lindblom & Hultquist: J. Bone & Joint Surg. 32:557)

These misdiagnoses led to the logical mismanagements shown in Table 3. On the few occasions where bed rest, heat and massage were prescribed *for the wrong reason*, some patients obtained temporary relief, but the physicians failed to re-evaluate their *patients' condition in the light of this favorable* response and to follow up with an appropriate support and advice as to the further use of the back.

Figure 1 shows a disk removed at autopsy These are the early changes mentioned previously, and this is the type of disk that can heal itself under conservative management.

Figure 2 shows how this healing under conservative management comes about. The *ruptured portion of the disk is invaded by* granulation tissue which digests the frag-



FIG. 2. Photomicrograph showing how healing takes place by invasion of digestive granulation tissue to absorb the broken portion of the disk. (Lindblom & Hultquist: J Bone & Joint Surg. 32:557)

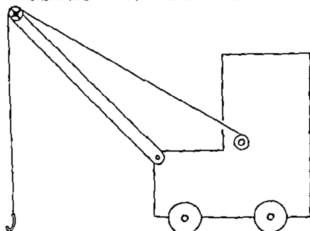


FIG 3. The power derrick has a solid lever arm with a fixed fulcrum and a power cable that will not tear under a load

mented material and in turn is replaced by scar tissue.

The following are some visual aids which have been helpful to me in explaining the back and its function to patients.

That the human back is not constructed like a power derrick and therefore should not be used as one will become obvious when Figures 3 and 4 are compared. Even the most obtuse patient will understand from this explanation that his back trouble is mechanical and that many times he can prevent attacks by being a little intelligent in its use. However, few patients realize how little musculature is available to extend their spine in comparison with that available to extend their knees. A brief perusal of Figures 5 and 6 will demonstrate to them why it is better to lift with the legs than with the back.

In conclusion, I wish to state that it is my firm belief that as physicians, and especially as orthopaedists, it is our responsibility to inform lay audiences whenever the

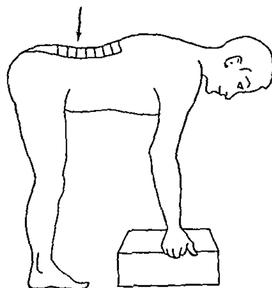


FIG 4. The human derrick (lifting with the extensors of the spine with the knees straight) has a multiple jointed rod as a lever arm, the lumbosacral disk as the fulcrum, and a set of power muscles which are short and fatigue easily.

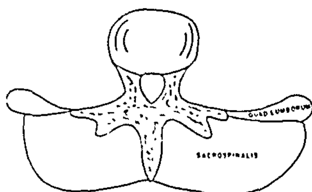



FIG. 5. Cross section, full size, third lumbar. A line drawing to indicate the volume of muscle available to extend the spine. Compare with Figure 6.

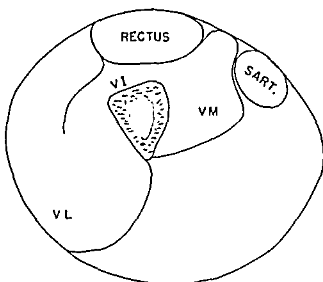



FIG. 6. Cross section, full size, mid-thigh. A line drawing to indicate the volume of muscle available to extend the knees (lifting with the legs). Obviously more muscle volume is available in this manner, and multiplied by 2, since there are 2 legs and only 1 back!

opportunity presents as to how disk injuries occur. We should not be surprised at the welcome receptiveness given to health suggestions, for most laymen are anxious to hear from their physicians on topics that will promote their general good. Failure to respond to invitations from lay groups misses a wonderful opportunity to cultivate the patient-physician relationship, still a subject of present-day interest. We orthopaedists can teach our fellow physicians at a staff or a county medical meeting, or give individual instruction by letter or demonstration on a patient in consultation. Fewer and fewer patients will seek care at the hands of the cultists if their physician is interested in giving competent back care.

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## Le Pathogenese de Lesiones de Disco Lumbar

*Summario in Interlingua*

Un revista de 4,000 consecutive casos de lesion de disco lumbar revela un definite correlation inter le typo del lesion e le pathologia del discos. In juvenes ancora crescente, le precoce exposition a labores rude e a certe typos de gymnastica se ha provate nocive. In adultos, certe formas de labor in que le dorso human es usate como

un derrick ha ledite un terrific numero de discos lumbar.

Es presentate al medicos plure recommendationes pro prevenir tal occurrentias o al minus pro tractar los promptemente a un tempore quando le correcte diagnose offere ancora melior prospectos de restablimento. Le valor de assignar selectivemente le un o le altere forma de labor es discutite con roentgenogrammas del spina dorsal como documentation additional.

Es *postulate* le *urgente desiderato de* plus adequate education del medico de fa lia qui vede al minus 67% de iste caso stadios non-avantiate e ergo ha le me opportunitate pro facer le correcte diagn e pro instituer le appropriate therapia. es clarmente un responsabilitate del or pedo in le scholas medical, in le clinica como consulente de su collegas de prac general.

# 15

# The Painful Coccyx

BECKETT HOWORTH, M.D., MED. SC.D.*

The word *coccyx* is derived from the Greek word for cuckoo because of the resemblance of the coccyx to the beak of this bird. The painful coccyx is commonly called coccygodynia. This word was proposed by Simpson in 1859, but it is the term for a symptom rather than a disease. The coccyx and its joints are subject to the same basic disorders as the musculoskeletal system in general. The coccyx may be affected by congenital deformity, fracture, infection or tumor, whereas its joints may be affected by sprain, arthritis or infection. Rarely is it the center of psychoneurotic attention. However, except for trauma and osteoarthritis, disorders of the coccyx are rare.

## HISTORICAL BACKGROUND

A careful, but not an exhaustive, search has been made of the literature pertaining to the coccyx. Apparently, the first mention of the painful coccyx was made in 1588 by Smet,[41] who said, "My wife has fallen backward and so injured the coccygeal bone that she cannot sit without pain, nor can she empty the bowel or the bladder, or cough without much distress." In 1862, Meek'ren[35] reported a dislocation of the coccyx that was reduced by manipulation on the third day with prompt and complete relief. In 1698, Gahrliep[22] mentioned that his father-in-law fell on icy steps and fractured the coccyx. He had great pain, "could get no passage of the bowels and became seriously ill on the fifth day, when he was advised to allow himself to be tumbled and pushed about by two robust women. By the jostling, the end of the os coccygeus, which was bent in, was restored," and he was relieved. In 1726, Petit[39] mentioned luxation of the coccyx with childbirth. He advised reduction of any luxation of the coccyx by manipulation. He reported a case of abscess of the coccyx, which he incised, but the patient died 5 months later. The diagnosis is not clear. In 1758, Pickett advised coccygectomy for persistent pain at the coccyx but reported no case. In 1827, Jacob[29] presented "an account of a remarkable production, resembling a tail, which was attached to the extremity of the vertebral column of a man" and mentioned another that had been "amputated by his father." He spoke also of "a family, several of whose members presented this remarkable variety of malformation."

Thus, as early as the seventeenth century, several types of painful coccyx had been seen and recorded. The emphasis in most of the subsequent reports has been on treatment.

In 1840, Blundell[6] described various lesions of the sacrococcygeal joint, such as inflammation, suppuration, ankylosis. He advised fracture of the ankylosed joint if necessary for delivery, removal of the coccyx for pain of unknown origin or for suppuration if other treatments failed. In 1844, Nott[37] said that the seat of pain was not always the seat of the disease, which might

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lead to error in diagnosis, and he reported an actual removal of the last two segments of the coccyx for neuralgia due to caries. In 1859, Simpson,[40] in an excellent paper that even today is timely, reviewed the literature and described the symptoms, the pathology and the treatment of fractures, dislocations and obstetric injury to the coccyx, reported 2 cases of teratoma, proposed the term *coccygodynia*, and advised subcutaneous stripping (rather than removal) of the coccyx for relief of persistent pain. Apparently this operation was not very successful, as it was not adopted by others.

In 1893, Darrah[13] reported 3 cases of caries of the coccyx. He stated that there were 23 cases in the literature and that 16 of 19 excisions had been successful; however, these were not proven cases. In 1903, Caubet[11] added 3 cases, and, in 1924, David[14] reviewed the subject and added 2 cases treated by coccygectomy. The latter stated that there had been only 1 report on the subject in the interval. In 1928, Blount[5] reviewed the literature of osteomyelitis of the coccyx, totaling 31 cases, but stated that many were not proven. He included the report of a case that was proved. It appears likely that in these various reports tuberculosis and other infections of the sacrum and the coccyx were confused and mixed together.

Coccygectomy was undertaken in earnest by the prominent proctologist Gant,[23] who, in 1923, reported 100 coccygectomies for fracture, dislocation, war injury, deformity, necrosis or periostitis but gave no follow-up or documentation. In his textbook he also gave an excellent chapter on lesions of the area of the coccyx, a chapter that curiously has been omitted from modern textbooks of proctology. Other authors were much more conservative in recommending coccygectomy. In 1924, Graff[24] reported coccygectomy during labor and, in 1928, Heckscher[26] discussed ossification of the joints as an obstacle to delivery. In 1937, Duncan,[19] in a good general article on the coccyx, reported 30 coccygectomies with full relief in 22; in the same year, Key[30] reported 15 coccygectomies with at least 12 very good results. Both these reports are well worth reading.

Massage of the painful saccrococcygeal area was advised in 1910 by Ely,[20] but this did not become popular until its vigorous advocacy by Thiele[44] in 1937. Other reports of good results with this treatment soon appeared: in 1937, by Hobart;[27] in 1939, by Foster;[21] in 1942, by Bourgouin;[8] in 1943, by Wilensky;[47] and in 1947, by Wilkinson.[48] Injection of the sacrococcygeal area with alcohol was advised in 1919 by Yeomans;[49] with procaine by Mandl[34] in 1929 and by Suermondt[42] in 1937; whereas in 1937 Waters[46] proposed both. Quinine and urea hydrochloride were preferred by Yodice[50] in 1932 and by Kleckner[31] in 1933. Radiotherapy was proposed by Baastrup[1] in 1915, who had used it in 15 cases with success in 10. In 1938, Beaton[4] suggested the possibility that coccygodynia is referred pain due to pressure of the piriformis muscle on the sciatic nerve. However, it seems clear that this is rarely the source of sciatic pain of any type. In 1951, Dittrich[15] suggested that the pain was associated with a degenerated fat nodule in the mid-sacral region and used procaine injection in this region. Certainly this would seem to be the cause of the pain in only an occasional case. It is of interest that there has been no other report of consequence concerning the painful coccyx in the past 20 years.

## ANATOMY

Usually, the coccyx consists of 4 bony segments (in about 80% of people), sometimes 3 or 5. In reality, the coccyx is an attenuated tail, but it is short and tucked forward underneath the skin. Rarely is a baby born with a coccyx *hyperextended* or long enough to project under the skin like a diminutive tail Although tails several inches long have been alluded to, no documented case has been located in the literature, and it is doubtful that a true human tail is pos-

sible embryologically. The coccyx of a woman is more exposed to injury than that of a man because her coccyx and lower sacrum are farther posterior, and the distance between the ischial tuberosities is greater.[19]

The coccygeal segments are rudimentary vertebrae, and there is a main mass or body with small transverse processes. The first segment is largest and resembles a sacral vertebra; it has definite transverse processes and proximal prolongations or cornuas that represent the superior articular processes. These processes are not always symmetric in size or in shape. There are no pedicles, laminae or spinous processes. The last segment is merely a small rounded nodule. (Figs. 1, 2 & 4)

The sacrococcygeal joint is a symphysis. There is a fibrous joint between the first and

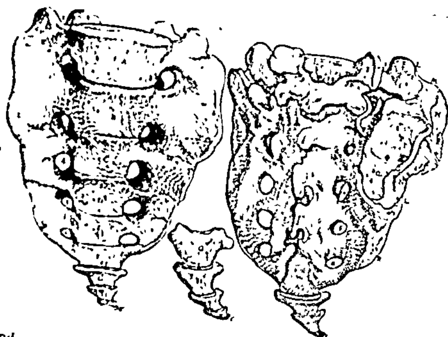


FIG. 1. Sacrum and coccyx, front and rear views, by Jan Calcar, from Vesalius' *De humani corporis fabrica*, published in 1543. There is a moderate deviation of the coccyx to one side. Although these figures were drawn more than 400 years ago, it would be difficult to improve upon their accuracy or their artistry today. It is interesting that the first sacral vertebra shown here is a transitional one, with features of a fifth lumbar vertebra

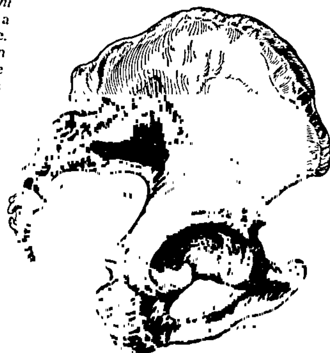


FIG. 2. Lateral view of right half of innominate bone, sacrum and coccyx. The segments of the coccyx are curved forward in line with the natural curve of the sacrum. In a normal or an erect sitting position the coccyx would be up out of reach of the seat, with no pressure against it (Ghormley, R. K. J.A.M.A. 101:1774)

Iliolumbar ligament
Supraspinal ligament
Short post. sacroiliac lig
Sacrospinous ligament
Superficial post. sacrococcygeal ligament
Sacrotuberous ligament

FIG. 3. Ligaments of the sacrum and the coccyx from behind. The posterior sacrococcygeal ligament consists of a long superficial and a short deep portion (Gray's Anatomy, ed. 24, Philadelphia, Lea & Febiger)

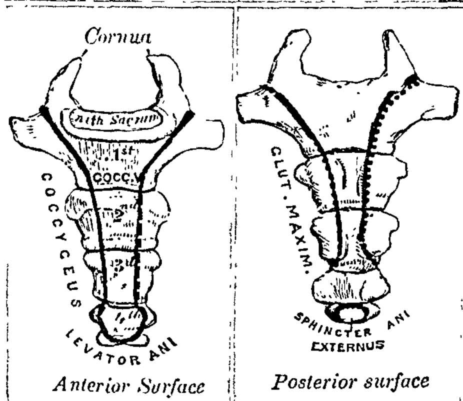



FIG 4. The coccyx, anterior and posterior surfaces. The first segment has transverse processes and cornua for articulation with the sacrum. The areas of muscle attachment are shown. (Gray's Anatomy, ed. 26, Philadelphia, Lea & Febiger)

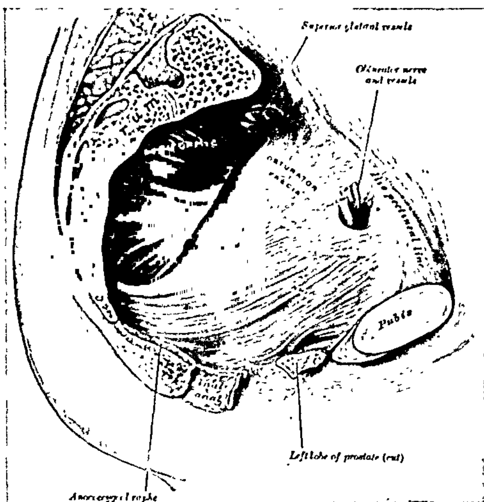



Fig. 5. Muscles of the coccyx as seen from within the pelvis The coccygeus extends from the spine of the ischium to the lateral border of the lower sacrum and the coccyx. The proximal portion of the levator ani extends from the ischial spine and the arcus tendineus to the lateral border of the last two segments of the coccyx and the median anococcygeal raphe. (Gray's Anatomy, ed. 26, Philadelphia, Lea & Febiger)

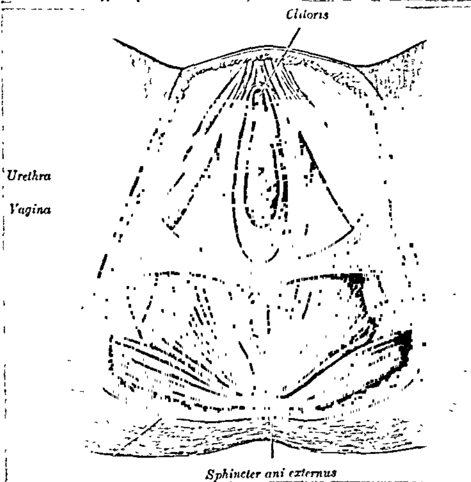



Fig. 6. Muscles of the coccyx from below The distal fibers of the gluteus maximus arise from the lateral surfaces of the coccyx The posterior fibers of the levator ani insert into the lateral border of the two distal coccygeal segments and the anococcygeal raphe. The sphincter ani externus surrounds the anus and has an attachment to the tip of the coccyx (Gray's Anatomy, ed 26, Philadelphia, Lea & Febiger)

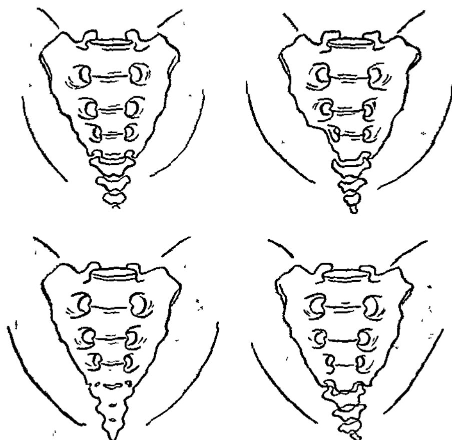


FIG. 7. Tracings from anteroposterior roentgenograms of sacrum and coccyx. (*Top, left*) Normal contours and relationships, joint spaces preserved. (*Top, right*) Asymmetry of last sacral and first coccygeal segments, with slight deviation to one side. (*Bottom, left*) Bony ankylosis of sacrococcygeal and coccygeal joints. (*Bottom, right*) Slight asymmetry of first coccygeal segment, moderate deviation of coccyx to one side.

the second coccygeal segments, and usually between the second and the third. The sacrum and the coccygeal segments are further held together by anterior, posterior and lateral ligaments (Fig. 3). In most cases, moderate motion is found between the sacrum and the coccyx, less between the coccygeal segments. The coccygeal segments tend in time to fuse together in the adult, earlier in men than in women.

In addition to its attachment to the sacrum, the coccyx is supported by certain muscles of the pelvic outlet The principal muscles of the coccyx are the distal fibers of the gluteus maximus extending from the lateral surface of the coccyx to the iliotibial band and the gluteal tuberosity of the femur, the coccygeus from the spine of the ischum to the lateral border of the lower sacrum and the upper coccyx, the iliococcygeus (a portion of the levator ani) that extends from the ischial spine and the arcus tendineus to the lateral border of the last 2 segments of the coccyx and the median anococcygeal raphe, and the sphincter ani externus that surrounds the anus and has an attachment to the tip of the coccyx (Figs. 5 & 6). All these muscles tend to pull the coccyx forward; when they act more strongly on one side, they pull it to that side. Essentially, there are no muscles extending the coccyx. Thus, the coccyx is always found to be flexed or curved forward, unless it has become ankylosed in the extended position

## ETIOLOGY AND PATHOLOGY

The coccyx varies in size, shape, direction and mobility in different individuals and at different ages. Normally, the coccyx is curved or angled forward by muscle pull, so that the individual is not conscious of its presence. In some people it deviates to one side; in others, it projects straight down, or even slightly backward (Fig. 7). In some individuals, motion of the coccyx is limited or absent, and if the coccyx has become stiff in the extended position its prominence is easily felt. When the coccyx is angled straight forward, the lower end of the sacrum is unduly prominent, especially in thin per-

sons. There may be a sharp angulation between the sacrum and the coccyx or between the first and the second coccygeal segments, with resulting bony prominence. Occasionally, the coccyx is free, "floating," or congenitally absent. Spina bifida of the lower sacrum is not uncommon.

The sitting posture of the individual has an important effect in the production of symptoms from a prominent coccyx or sacrum. When a person sits erect with a normal lumbar lordosis and the weight is supported on the thighs, the coccyx does not reach the seat. However, when the individual flattens or reverses the lumbar curve and slumps forward, the coccyx drops and reaches the seat, causing painful pressure. A jolt in this position may cause injury to one of the joints of the coccyx. A fall on the buttocks or astride is a frequent source of sprain of the coccyx, occasionally of fracture or dislocation. Traumatic osteoarthritis follows repeated injuries to its joints. Injury to the joints of the coccyx during childbirth leads to fibrosis and reduced motion, which in later deliveries may require fracture of the ankylosis, leading eventually to bony ankylosis, sometimes in an extended or a deformed position (Fig. 12).

The pathologic findings in injuries to the coccyx or its joints are comparable with those seen in similar lesions in other areas; e.g., congenital deformities, fractures, dislocations, osteoarthritis.

A coccyx sharply angulated forward may cause damage to the rectum, even perforation or ulceration. Such a coccyx may damage the local soft tissues, or even the head



FIG 8. Anteroposterior roentgenograms of lower sacrum and coccyx at such an angle as to bring the coccyx above the symphysis for better visualization. (*Left*) Normal lower sacrum and coccyx, almost symmetric, with healthy joints. (*Right*) Moderate asymmetry of lower sacrum and coccyx with partial ankylosis of sacrococcygeal joint.

of the infant, during delivery.[23] Infection of the coccyx is uncommon, but tuberculosis and osteomyelitis have been reported. Primary tumors of the coccyx are rare, but giant cell tumor has been reported;[33] sarcoma of the sacrum is seen occasionally; metastatic carcinoma of the lower sacrum and coccyx is less rare (Fig. 17). Chordoma of the sacrococcygeal region is rare in the experience of any one surgeon, but the Mayo Clinic reports 48 cases[12] (Fig. 16). Glomus tumor of Luschka's body, an arteriovenous plexus anterior to the lower sacrum, is also rare. Teratoma and meningioma of the coccygeal region are seen usually in infants and generally are large enough to be readily recognized.[40]

## SYMPTOMS

The usual symptom related to the coccyx is pain. The pain is present commonly on sitting, often relieved by standing or lying down. When the pain is acute, it may be aggravated by lying on the back, by walking or stooping, or by any contraction of the muscles or movement of the coccyx. The pain is worse when sitting upon a hard seat or sitting with the lower back slumped. The pain is increased further by sitting in a moving vehicle or riding on horseback.

The pain begins suddenly with a definite injury in many cases, but it may start gradually with no obvious cause. The pain tends to persist with faulty sitting posture and with much sitting. The pain may be worse with coughing or straining at stool, especially with constipation or with rectal or vaginal disease. The vast majority of individuals with a painful coccyx are women, and the usual age period is between 30 and 50 years, but even a child may have persistent pain in

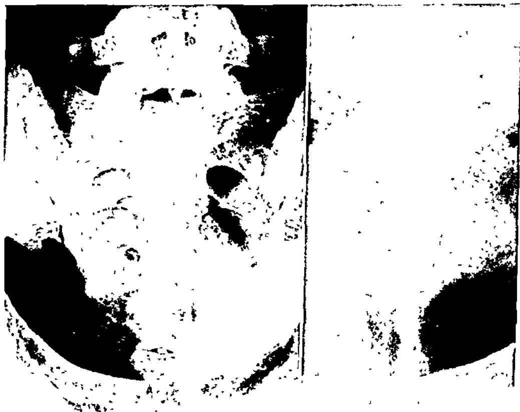


FIG 9 Anteroposterior roent f sacrum and *eft)* Calcification in center of sacroco ysis. The join in, with condensation of adjacent surf c degenerative ich in time will lead to bony ankylosi (*Right*) An ith bony ankylosis of sacrococcygeal a ints.



FIG 10. Lateral roentgenograms of sacrum and coccyx of two children. (*Left*) A 10-year-old girl. The sacral segments are not yet fused, and only the first coccygeal segment is ossified The curve of the sacrum and the coccyx is normal. This child had pain of several months' duration at the coccyx following a fall on the buttocks, with tenderness and pain on motion. The pain was relieved in a short time by teaching her to keep the sitting weight off the coccyx, to apply local heat and to do gluteal muscle exercises. (*Right*) A 13-year-old girl. The sacral segments are nearly fused, but the distal sacral epiphysis is yet visible. The coccyx is angulated forward sharply between the first and the second segments. The girl had persistent pain and tenderness, relieved by protection and gluteal exercises.

the coccyx after a direct injury (Fig. 10). Coccygeal pain may be produced or aggravated by delivery of a baby.

## SIGNS

The principal signs of lesions of the coccyx are *localized* tenderness and pain on motion. The tenderness is usually at the sacrococcygeal joint, less often at the joint between the first and the second segments. Pain is produced by motion of the affected joint, either by pressing the coccyx forward or by moving it back and forth with one finger in the rectum. Swelling, inflammatory signs, muscle spasm and even ecchymosis may be present *after* recent injury. The symptoms and signs are proportional to the severity of the injury. There is a tendency to sit on one buttock or to support the weight of the trunk with the arms to relieve pain. If there is a fracture, the signs are localized to the bone rather than to the joint, unless the joint, too, is involved. Displacement of the fragments may sometimes be felt, but this is not usual Deviation of the coccyx forward or to one side is not due necessarily to a fracture or a dislocation and may have been present long before the injury. Definite displacement of a joint is indicative of dislocation, whereas displacement of the fragments may be palpated with displaced fracture of the lower sacrum or coccyx.

Rectal examination should always be done in order to palpate the anterior surface of the coccyx, the lower sacrum and the adjacent muscles, as well as to rule out or to study a rectal lesion. Pelvic examination sometimes is advisable.

## ROENTGENOGRAMS

Anteroposterior and lateral roentgenograms of the lower sacrum and coccyx are advisable in all cases of coccygeal pain in order to demonstrate anatomic variations as well as the possibility of a pathologic process (Figs. 8 & 9). The rectum should be emptied beforehand, preferably with a laxative or an enema. Congenital anomaly or asymmetry of the coccyx is not uncommon and is usually without clinical significance (Fig. 8). Forward or lateral deviations of the coccyx should be distinguished from displacements due to fracture or dislocation (Figs. 1 & 10). Fracture, when present, usually extends transversely or obliquely across the lower sacrum or the first coccygeal segment (Fig. 14). Dislocation is rare as a traumatic lesion, but, when present, it is seen more often at the sacrococcygeal joint (Fig. 15). Osteoarthritis of the sacrococcygeal joint is sometimes recognizable in the roentgenograms (Fig. 11). Bone destruction due to infection or tumor is seen occasionally.

## DIAGNOSIS

An acute injury, with acute local symptoms and signs, is usually indicative of a sprain, occasionally of a fracture, and roentgenographic confirmation is advisable. Chronic pain in the region of the coccyx is

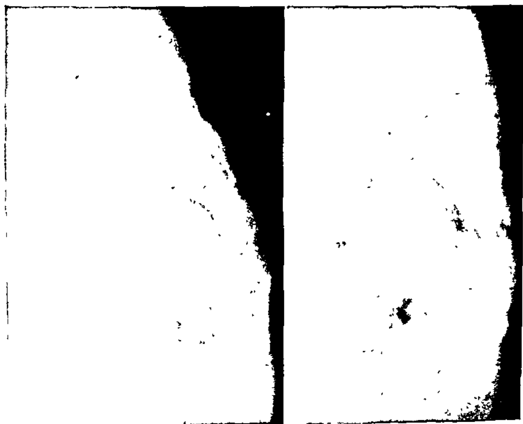


Fig 11 A 45-year-old woman. There is ankylosis of the sacrococcygeal and the coccygeal joints, largely bony, and the coccyx points straight down, due partly to the increased lumbar lordosis. The coccyx, stiff at childbirth, had been freed by manipulation This was followed by pain and spasm for several weeks Sitting pain had persisted, requiring eventual removal of the coccyx, which brought relief (*Right*) A 43-year-old woman The coccyx is curved forward, but there is moderate osteoarthritis of the sacrococcygeal joint with fibrous ankylosis. The persistent pain and tenderness were largely relieved by a proper sitting posture and physical therapy.

usually due to traumatic osteoarthritis but sometimes follows an acute sprain or fracture. Pain due merely to local irritation from prolonged sitting with poor posture is less common.

Low-back pain sometimes radiates into the sacral or the coccygeal region, but the signs are localized to the low back rather than the coccyx (Fig. 13). Dittrich[15] reports fat nodules dorsal to the lower sacrum as a source of coccygeal pain, but this has not been confirmed by others.

Lesions of the anus and the rectum, as well as perirectal abscess, fistula and pilonidal cyst, are distinguished by careful external, rectal and proctoscopic examinations. A glomus tumor of Luschka's body or other tumor may be felt on rectal examination, and roentgenograms may be confirmatory if there is bone destruction.

Tumor of the cauda equina may cause pain or numbness, or both, in the region of the coccyx, but the symptoms and signs are more widespread. Differentiation requires careful examination of the low back and the rectum as well. Neurologic opinion may also be desirable.

Spasm of the muscles of the coccyx and vaginismus have been reported as sources of coccygeal pain (Ely,[20] Thiele[11] and others). Often, pain in the coccygeal region has been attributed to psychoneurosis, especially since it occurs so preponderantly in women. However, it is important to distinguish those who have a definite local lesion of the coccyx or its joints with exaggerated symptoms from those who have no local lesion. Persistent coccygeal pain in a sensitive woman may easily lead to a mistaken diagnosis of a psychoneurosis.

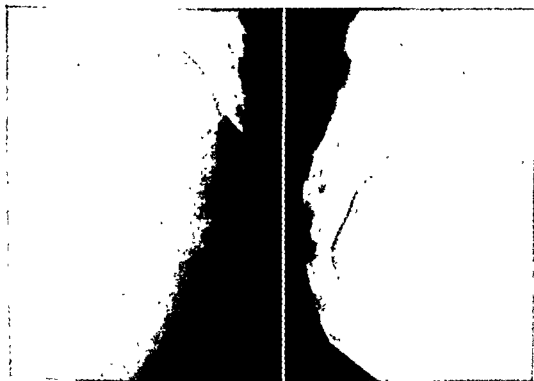


FIG. 12. A 50-year-old woman. The coccyx is sharply angulated forward through the first segment, deformed apparently by a fracture associated with a severe fall on the buttocks. There is bony ankylosis of the sacrococcygeal joint. Partial relief was obtained by a proper sitting posture and appropriate physical therapy. (*Right*) A 52-year-old woman. There had been difficulty with the coccyx on two deliveries, as well as several falls on the buttocks. The coccyx is curved forward, with bony ankylosis of all joints. Relief of persistent pain and tenderness was obtained by coccygectomy.

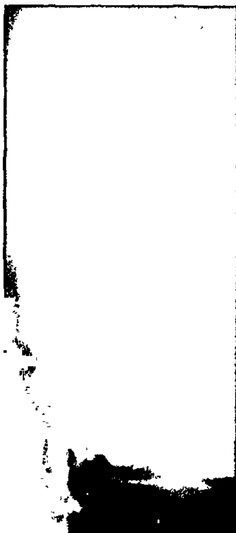
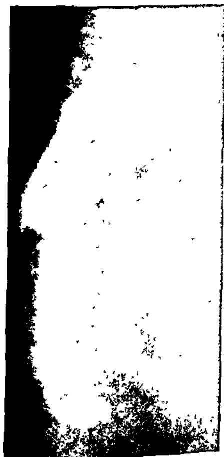


Fig. 13. *A 48-year-old woman. The lumbosacral disk* is thin, with degenerative changes. The lumbar spine is erect and flat, being almost in vertical alignment with the sacrum. Thus, the coccyx is directed downward, with persistent pain and tenderness. In this instance the lumbosacral abnormality was largely responsible for the coccygeal symptoms. However, removal of the coccyx led to relief. The lumbosacral joint also required treatment

Fig. 14. Fracture of the lower sacrum, transverse, comminuted, with moderate forward displacement of the lower fragment, and slight forward angulation. Such a displacement should be reduced by manipulation if possible. There is also a moderate forward angulation of the lower coccyx that had existed previously.



## TREATMENT

Recent dislocation or fracture displacement of the coccyx should be corrected by digital manipulation by rectum, using anesthesia if necessary. Sometimes the manipulation has to be repeated because of persistent or recurrent displacement. Digital fracture of the coccyx for a stiff sacrococcygeal joint is sometimes advisable at the time of delivery.

The essence of treatment of acute or chronic traumatic disorders of the coccyx is to keep weight off the coccyx until the *joint has had time to heal. This is done by* using an upright, properly fitting chair with a properly shaped and firm seat, and by using the upright sitting posture with the weight mostly on the thighs. A pad or a small pillow may be placed in the small of the back to aid in maintaining this position. A sponge-rubber cushion may be desirable *for the seat.* The pain may be relieved by hot sitz baths, hot packs or a hot-water bag, or rectal douches. External massage is sometimes helpful. Massage per rectum of

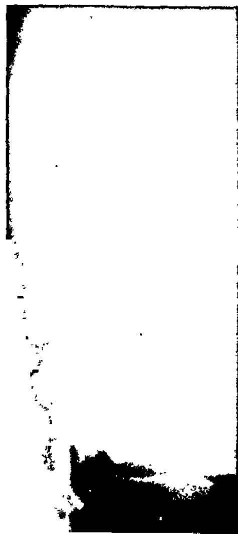


FIG. 13. A 48-year-old woman. The lumbosacral disk is thin, with degenerative changes. The lumbar spine is erect and flat, being almost in vertical alignment with the sacrum. Thus, the coccyx is directed downward, with persistent pain and tenderness. In this instance the lumbosacral abnormality was largely responsible for the coccygeal symptoms. However, removal of the coccyx led to relief. The lumbosacral joint also required treatment.

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FIG. 15. (*Left*) Posterior subluxation of the coccyx, with osteoarthritis in sacrococcygeal joint, restricted painful motion, relieved by coccygectomy. (*Right*) Sharp forward angulation at second and third coccygeal joints. There was poor motion at the sacrococcygeal joint, with pain, and tenderness at the kyphos. A good result was obtained by removal of the coccyx. (Key & Conwell: Fractures, Dislocations and Sprains, ed. 6, St. Louis, Mosby)

the sacrococcygeal region and the surrounding muscles is beneficial when the muscles are in spasm and tender, especially after childbirth. Massage is best given 2 or 3 times weekly for 3 or 4 weeks. Specific exercises for improving the tone of the muscles of the buttocks are helpful. Weight reduction is advisable in the presence of obesity.

Radiotherapy, as well as various injections into the sacrococcygeal area or nerves, has been used in the past, with conflicting reports as to the results of such treatment. However, local but careful and precise injection of hydrocortisone with 1 per cent procaine or lidocaine has proven to be more successful than any of these earlier treatments and often gives sufficient relief to obviate surgery.

## COCCYGECTOMY

This operation was first suggested by Pickett in 1758, again by Blundell[6] in 1840,

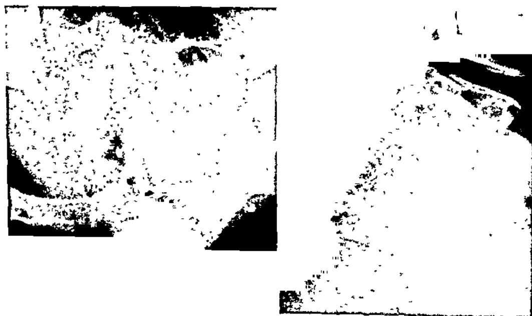


FIG. 16. (*Left & right*) Chordoma of the sacrum. Much of the lower half of the sacrum has been destroyed by this tumor, leaving only strands of trabeculae inside the cortical outline of the bone. When such a tumor in this location is small, the symptoms may be confused with those of traumatic arthritis of the sacrococcygeal joint.



FIG. 17. This patient had persistent pain in the sacrococcygeal region, worse at night. There is a spotty area of increased density and irregular outline near the center of the sacrum, a sarcoma. Again, confusion as to diagnosis may arise early in the course of the disease, especially if good roentgenograms are not available

and first done in the United States by Nott[37] in 1844. Removal of the coccyx is advisable when there are persistent, localized pain and tenderness unrelieved by nonoperative treatment and when the diagnosis is clear. Coccygectomy is not a panacea for pain in the coccyx and should not be used unless there is a clear indication, such as a stiff coccyx in a badly deformed position, diseased bone, a removable tumor or persistent unrelieved localized pain. The operation should not be done in the week preceding or during the menses.

The operation requires meticulous care in the dissection, and it should be remembered that what is left behind is more important than what is removed. Ragged dissection, hematoma, bony spicules and bad scars are not conducive to good results. We have seen at least one failure due to a long spike of bone that was left projecting downward into the area of dissection.

Coccygectomy is best done with the patient prone, hips and knees flexed moderately, knees separated. The skin incision should be to the side of the mid-line, as it will heal better. Posterior dissection should adhere closely to the periosteum, and the lateral dissection should follow carefully the contours of the bone, including the transverse processes. Usually the coccyx is disarticulated from the sacrum and pulled back gently, and the periosteum is stripped carefully from its anterior surface. Sometimes it is helpful to disarticulate other portions of the coccyx in the process of removal, but it is important not to lose sight of the terminal segment. Usually it is advisable to remove the distal narrow portion of the sacrum, about ¼ inch above the joint, leaving a smooth, rounded stump. More should not be removed because of the danger of weakening the rectal floor. The entire dissection should be neat and clean, and bleeding should be controlled carefully. The periosteum and the ligaments are sutured carefully to close the dead space, particular attention being paid to avoid puncturing the rectum. The subcutaneous tissue and the skin are closed separately. A thin dressing is applied and may be sealed with collodion to prevent contamination. This may be covered with additional dressing, held with a T-binder and changed as indicated

A constipating diet and pill of opium or morphine are given commonly for 2 or 3 days after the operation, followed by an enema. However, it seems preferable, instead, to give Thalamyd (phthalylsulfacetamide) 2 0 Gm. 3 times daily for 3 days before operation and 3 days after operation to serve as an intestinal antiseptic, and to maintain normal bowel function and avoid constipation. Gluteal exercises are resumed a few days later. The patient may be ambu-

lant as soon as this is comfortable. The sutures are removed in 7 to 10 days, and sitting is begun gradually.

The results of the operation are very good if the cases are chosen carefully and skillful technic is used. Gant,[23] in 1923, reported good results in 90 per cent of 100 cases but did not present a critical analysis. In 1937, Duncan[19] reported good results in 22 of 30 cases, Key[30] in 12 of 14 operations. The author has had good or excellent results in 11 coccygectomy cases with long follow-up.

## PROPHYLAXIS

Chronic strain of the sacrococcygeal joint may be avoided by the use of proper seating and good sitting posture, by the maintenance of good gluteal muscle tone and the avoidance of obesity. Usually, falls on the coccyx may be avoided by the elimination of obvious dangers such as loose rugs and polished floors, the avoidance of high heels, and keeping the weight forward with the body in a slight crouch when walking on slippery surfaces (dynamic posture).[28]

## CONCLUSIONS

An understanding of the anatomy of the coccyx and its joints is helpful in evaluating its disorders. Injury and disease of the coccyx are subject to accurate diagnosis, as are any other bone and joint disorders. Lesions of the coccyx are comparable with those of other parts of the skeletal system. Traumatic osteoarthritis and sprain are the most common disorders of the coccygeal joints. True psychoneurosis related to the coccyx is rare.

Pain in the coccyx can be relieved usually by removal of the weight from this area in sitting, aided by physical therapy.

Persistent pain in the coccyx may be relieved by coccygectomy, but the operation should be chosen carefully and performed well.

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## Le Coccyx Dolorose

*Summario in Interlingua*

Coccygodynia non es un diagnose sed un symptoma. Ab le puncto de vista diagnostic, dolores coccyge debe esser evalutate secundo le mesme principios como lesiones e morbos de non importa qual altere parte del systema musculoskeletic. Le coccyx e su articulationes suffre, a generalmente parlar, le mesme disordines como omne altere parte de ille systema.

Es presentate un revista del litteratura

relative al coccyx dolorose a partir de 1588 usque al tempore presente. Es describite brevemente le anatomia del coccyx, su ligamentos, articulationes, e musculos. Le etiologia del varie affectiones del coccyx e su articulationes es discutite. Le tableau clinic correspondente al varie problemas del coccyx es presentate, incluse le constatationes roentgenographic e le diagnoses differential.

Le tractamento de varie typos de dolores del coccyx es reportate e discutite, incluse le tractamento del typo usual incontrate in jectos sedentari in le absentia de tra specific, del typo incontrate post parto, fracturas e dislocationes. Varie forma injection local, radiotherapia, e therapia tural e physic es evalutate.

Coccygectomia esseva proponite in 1 e primo effectuate in iste pais per Not 1844. Le indicationes pro coccygectomi discutite. Le technica, le cura post-op tori, e le resultatos es reportate.

# 16

# An Operation for the Correction of Metatarsus Primus Varus Applicable to Both Children and Adults

GUILLERMO PARRA, M.D.,* AND DUNCAN C. MCKEEVER, M.D.†

*Adventure on, for from the littlest clue*
*Has come whatever worth man ever knew;*
*The next to lighten all men may be you.*‡

Many surgical procedures have been devised for the correction of metatarsus primus varus and so-called splay foot. Those involving the bone have a long convalescence, and most of them are not applicable to small children, who may have this condition to such a degree that hallux valgus with exostosis occurs at a very early age or is certain to develop later in life Sling procedures are passive in nature, and the correction may not be permanent.

The procedure reported here was first devised for and used on children. As the technic became more standardized, it was used for young adults and certain selected older adults. It does not replace or supersede arthrodesis of the first metatarsophalangeal joint, which continues to be used as the operation of choice in hallux rigidus and in those cases of metatarsus primus varus having marked hallux valgus deformity with large exostoses and subluxation. To date, it has been used more often in those children having severe metatarsus primus varus deformity and in those adults who have flexible feet without actual metatarsophalangeal subluxation.

Basically, the procedure is a modification of the McBride technic of transfer of the conjoined tendon; however, the McBride operation decreases the mechanical advantage and functional strength of the active muscles by bringing the origin and the insertion closer together. This operative procedure increases the mechanical advantage of the muscle by further separating its origin and insertion and converting the action into a more direct force for correction of the metatarsus primus varus.

The "littlest clue" from which this procedure was developed was given one of the authors by the other in the form of a skeletal foot with some rubber bands pulled across between the origin of the oblique and the transverse heads of the adductor hallucis and a new point of insertion on the medial side of the head of the first metatarsal (Fig. 1). The increased mechanical advantage resulting from more direct action and the greater range of potential movement were immediately apparent.

To date, the authors have carried out this operative procedure on 40 cases with cor-

* Mexico City

† Houston, Tex

‡ Masefield. Poems, "The Ending," New York, Macmillan, 1935 (copyright Barbara Zucker).



Fig. 1. "The littlest clue."



Fig. 2. Tendon separated from sesamoid and insertion.

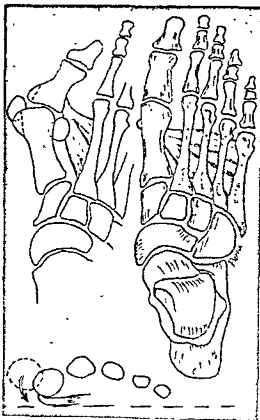


Fig. 3. Showing the altered mechanics of muscle action after removal of exostosis and transplant of tendon.

rection of the deformity and without complication. The technic now is fairly well standardized, and no change has been made recently.

THE TECHNIC

A 1½- to 2-inch linear incision is made from a point near the base of the proximal phalanx proximally along the superior lateral border of the shaft of the first metatarsal. Through this incision the adductor hallucis tendon is isolated and tenotomized at its insertion. It is separated from the lateral sesamoid, which usually is removed in adults and left intact in children. The tendon is separated from all surrounding structures back to the point at which the muscle body becomes visible (Fig. 2).

A second incision is made on the medial side of the foot toward the plantar surface. The skin is reflected. The bursa over the bunion is removed, the periosteum being incised immediately above the level of the middle of the medial surface of the first metatarsal head. The periosteum is reflected, and the exostosis is removed with a motor saw. The small opposing exostosis, usually present on the base of the proximal phalanx,

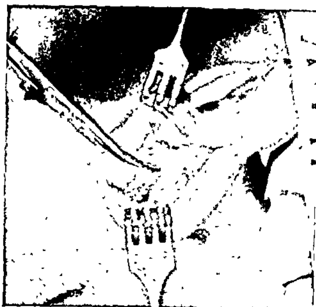


FIG. 4 Illustrating tendon passed under the neck of the metatarsal, ready for insertion.

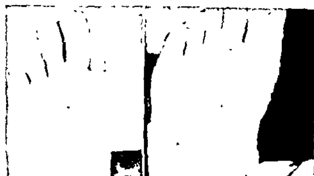


FIG. 5. Adult before operation (*left*) and after operation (*right*).

is also removed (Fig. 3). An extraperiosteal opening ½ to ⅝ inch in width is made beneath the neck of the metatarsal. The tendon is drawn through this channel and dorsally across the cancellous surface of the metatarsal head (Fig. 4). It is attached with interrupted stainless-steel sutures under the



FIG. 6. A 12-year-old boy before operation (*left*) and after operation (*right*).

edge of the capsule and periosteum of the head of the metatarsal near the extensor hallucis longus tendon. The capsule is closed over it. Both incisions then are closed, and a pressure dressing is applied.

Tension on the pressure dressing is released later the same day, and the following day a smaller dressing is applied. Within 48 hours, a single strip of gauze is applied over each incision and 4 or 5 turns of 2-inch adhesive passed round the forefoot. The patient then is ready for full weight-bearing in a cutout shoe. The sutures are removed in 2 weeks, and the patient is able to be more or less normally active. The 4 or 5 layers of 2-inch adhesive round the forefoot are continued for 5 weeks.

The authors consider that this procedure is sufficiently satisfactory to warrant its use for the correction of splay foot and metatarsus primus varus in children. It is also applicable to most cases of hallux valgus in adults (Fig. 5). A boy, aged 12, who underwent operation is shown in Figure 6.

## Un Operation pro Corriger Vare Metatarso Prime, Applicabile a Patientes Pediatric e Adulte

*Summario in Interlingua*

Il existe numerose technicas chirurgic pro le correction de vare metatarso prime e pede plan in juvenes e adultos. Plures require longe periodos de reconvalescentia; alteres non es permanente. Le technica describite in le presente articulo esseva elaborate originalmente pro juvenes. Al initio, illo esseva usate in casos juvenil solmente, sed gradual-

mente illo esseva applicate a juvene adultos e a seligite patientes de etates plus avantiate. Hodie illo es le operation de election in casos de halluce valge con vare metatarso prime, proviste que le articulation metatarso phalangee es ancora intacte.

In su principio le technica es un modification de illo de McBride. In illo, le transplantation del tendine conjuncte es manipulate de maniera a augmentar le fortia functional e le avantages mechanic del musculos in transposition. Figura 1 demonstra le avantage mechanic que resulta ab le action plus directe e le plus grande libertate del movimentos potential del musculo.

Le operation ha essite effectuate in plus que 40 casos, con le resultato de un correction del deformitate e sin le occurrentia de complicationes. Le technica es le sequente:

Un incision de inter 1½ e 2 pollices de longor es facite dorsalmente inter le prime e le secunde metatarsal. Via iste incision le tendine del adductor hallucal es isolate e tenotomisate al sito de su insertion. Illo es separate ab le sesamoide le qual es usualmente ablationate. Le tendine es separate ab omne le structuras circumjacente usque a un puncto ubi le corpore del musculo deveni visibile (figura 2).

Via un incision al latere medial del pede, le bursa e le exostose es removite, e similemente le exostose opposite—si illo existe—es removite ab le base del phalange proximal (figura 3).

Un tunnel extraperiostee es facite sub le cervice del metatarsal. Le tendine conjuncte es tirate a transverso le tunnel, super le area nude del osso ubi le exostose esseva excidite. Le tendine es suturate firmemente a in le capsula proxime al vaina del tendine del longe extensor hallucal. Le incision es claudite e un bandage de pression es applicate.

Plus tarde le mesme die, le tension del bandage es relaxate. Intra 48 horas un leve bandage de gaza e 4 o 5 circumvolutiones de banda adhesive de un largor de 2 pollices es applicate al parte anterior del pede. Alora le patiente es preste a supportar su peso. Le suturas es eliminate in 2 septimanas.

Le circuitos de banda adhesive al parte anterior del pede es mantenite durante 5 septimanas.

Como jam indicate, le operation ha essite effectuate in plus que 40 casos, con correction del deformitate e nulle complicationes. Illo es considerate como satis promittente pro justificar su uso in le correction de pede plan, vare metatarso prime in juvenes e etiam in le correction del majoritate del casos de halluce valge in adultos.

Figuras 5A e B e 6A e B es illustrationes pre-operatori e post-operatori.

# 17

# Functional and Cosmetic Correction of Metatarsus Latus (Splay Foot)

JOSEPH EDMUND BROWN, M.D.*

The woman of today is a victim of fashion, and particularly so as regards the foot. No harm comes from the unnatural compression or shifting of soft tissue to meet the demands of a Dior creation, but the bony foot rebels when it is jammed into a container—loosely referred to as footwear—much too small to accommodate it (Fig. 1).

The rebellion of the foot to abnormal pressure manifests itself in bunion formation, the accentuation or the development of the true hallux valgus deformity, corns on the dorsal surface of the lesser toes, plantar calluses, bunionettes, intradigital neuromata and metatarsalgia.

The orthopaedist dealing with any of the above painful subjective and objective manifestations of abnormal compression and pressure areas readily recognizes a simple solution; i.e., make the shoe fit the foot. However, such simple but sound advice goes unheeded because of the dictates of style; hence the foot suffers

A comely woman with a metatarsus latus or splay foot is usually an uncompromising and unhappy combination. Painful feet render the combination all the more complex Unquestionably, palliative measures directed toward the relief of the pressure phenomena do give the abnormally compressed foot temporary relief Excision of proliferative bone formation or the removal of adventitial bursa or neuromata may eliminate temporarily the presenting complaint of pain or discomfort but offers no true solution of the basic cause of these pressure phenomena.

A number of years ago, when confronted with an uncompromising matron with splay feet who had not obtained relief of symptoms following previous excision of intradigital neuromata and exostoses of the proximal phalanges, it occurred to the author that the only logical approach was to reduce the width of the forefoot. The rationalization of the proposed surgery was explained to the patient. In the spring of 1948 an operation was performed, the fifth toes along with the distal two thirds of the adjacent metatarsal shafts being removed. This woman was proud of her narrow feet. Unfortunately, she was lost to follow-up study, and her roentgenograms are no longer available, so that her particular case is included in this report only for historical background.

## THE OPERATION

An incision on the dorsum of the foot begins at the web space between the fourth and the fifth toes, and extends proximally to the prominence of the proximal end of the fifth metatarsal. A second incision begins in the web space and extends along the plantar aspect of the foot, crossing directly over the head of the fifth metatarsal and then curving

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gently and obliquely toward the proximal end of the first incision (Fig. 2, *top*). The island of skin is made narrow intentionally to allow for the necessary revamping for approximation in the final wound closure.

The ligaments between the metatarsal heads are cut. The intradigital nerves are inspected. If a neuroma is present, it is removed. The long extensor and flexor tendons are severed. The interosseous structures are divided in the mid-line to preserve a soft-tissue padding for the lateral aspect of the foot. The tendinous insertions of the peroneus tertius and brevis are reflected from the proximal end of the fifth metatarsal, and the bone is shelled out and disarticulated from the adjacent fourth metatarsal and the cuboid. The insertions are sutured to the remaining capsule and adjacent soft (surrounding) tissue.

The muscle and the fascial planes are approximated, and any abnormal *bulge* is excised. Skin hooks placed in the plantar flap reflect the *excess* over the dorsal skin edge. This surplus then is removed, resulting in a straight incisional line distinctly on the dorsum of the foot (Fig. 2, *bottom*).

A compression dressing is applied, and the same procedure is performed on the opposite foot. Ambulation is allowed according to the patient's tolerance.




FIG. 1. Roentgenogram of a young woman's foot bearing weight in and out of a conventional "pump" shoe. The natural width of the forefoot is compressed ⅞ inch to conform to the shoe.

## CASE REPORTS

Case 1. In May, 1954, a 24-year-old female with a
seen.
but a

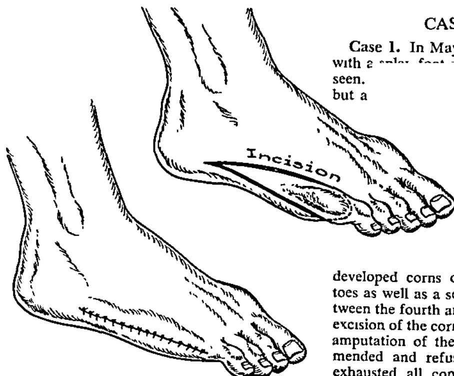



FIG. 2. (*Top*) The incision employed (*Bottom*) The incisional wound of the narrowed foot is on the dorsal aspect and not subject to pressure.

developed corns on the dorsum of the lesser toes as well as a soft corn in the web space between the fourth and the fifth toes. Arthrodesis, excision of the corns, removal of bone spurs and amputation of the fifth toes had been recommended and refused. She had, she thought, exhausted all conservative measures and ap-

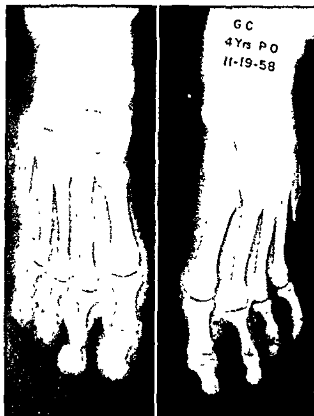



FIG 3. Roentgenograms of feet bearing weight 4 years after operation.

proaches and had literally *a closet full of shoes.* The thought of a narrow foot appealed to the patient.

On August 14, 1954, the fifth toes and most of the adjacent metatarsal shafts were excised. When this patient was last seen—November 19, 1958 (Figs. 3 & 4)—she reported that she did not have any pressure areas and was pleased to think that she was able to conform to fashion trends without foot discomfort. Her shoe size had been reduced from 9B to 9AAA

Case 2. On January 8, 1958, a 42-year-old registered nurse was seen with painful feet that had reached the point of almost incapacitating her. She had had endless chiropody treatment, and it had been recommended that she have intradigital neuromata excised from the web space between the fourth and the fifth toes.

Clinical evaluation revealed a typical splay foot (Fig. 5). The patient had a tendency toward a hallux valgus formation with areas of pressure evident in the region of both prominent first metatarsal heads. There were also bunionette formations on both fifth toes, and there was distinct tenderness in the web between the fourth and the fifth toes of both feet. This woman, married to an executive, had definite social obligations, and she admitted frankly that she was not ready to compromise in regard to her footwear.

It was explained to this patient that, in all probability, the multiple mechanical features involved would not respond to anything short of narrowing down the forefoot.

On January 21, 1958, both fifth rays of the [illegible]

[illegible] for her housework, and even then did not have foot comfort. Since surgery she has not made



FIG. 4. Weight-bearing photographs 4 years after operation. Note the bulges in the region of the proximal ends of the metatarsals.

3 5. Preoperative roentgenograms.



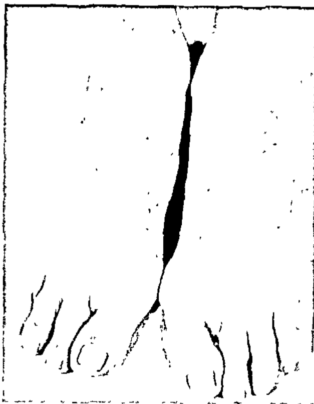


FIG. 7. Weight-bearing photographs 10 months after operation.

any complaints referable to the great toe, and she has not had any other signs of a compression-pressure phenomenon (Fig. 7) even when wearing high heels.

## SUMMARY

A method of dealing with a metatarsus latus or splay foot in an uncompromising patient is reported. It consists of narrowing down the width of the forefoot by excision of the fifth ray. In earlier cases, the proximal end of the fifth metatarsal was not removed. In subsequent cases, the entire fifth metatarsal shaft was excised, resulting in a better cosmetic appearance (compare Figs. 4-7).

The surgical correction is, indeed, cosmetic in one respect but functional in another, insofar as it does eliminate pressure phenomena and allows the patient to be *stylish,* yet comfortable.

The operative procedure has been performed in 4 carefully selected cases. It is not intended that this procedure become as popular as rhinoplasty, in spite of the fact that, in all probability, the indications equally justify it. The results have been sufficiently gratifying to warrant this report.

## Correction Functional e Cosmetic de Metatarso Late

*Summario in Interlingua*

Un femina que obedi al dictatos del moda in su calceatura es un candidata pro le sequente phenomenos de pression anormal si illa ha pedes plan, a saper, formation de tuberes verrucose, accentuation o disveloppamento de ver halluce valge, callositates, callos plantar, verrucositates minor, neuromas intradigital, e metatarsalgia.

Mesuras palliative es certo capace a alleviar tal phenomenos temporarimente, sed si le uso de anormalmente micre calceos es continuate, etiam le pression e su effectos va continuar a varie grados e in varie combinationes.

In 1948 le idea occurreva al autor del presente articulo que in le caso del femina qui non es preste a acceptar ulle compromisso le sol solution logic consisterea in reducer le largor del parte anterior del pede. Pro attinger iste objectivo, il esseva proponite remover le quinte digito pedal e le adjacente osso metatarsal.

Le technica chirurgic es illustrate in le accompaniante diagrammas. Le morbiditate que resulta del intervention es practicamente nulle. Le resultatos functional e cosmetic es extrememente gratificante.

In quatro cautemente seligite casos de metatarso late o de pede plan, excision del quinte radio esseva effectuate sin complication adverse, con le resultato del elimination de dolorose phenomenos de pression causate per le compression anormal del stricte calceos de moda que es infligite al feminas de nostre dies e que illas accepta.

# 18

# Parosteal Osteogenic Sarcoma

## Treatment by Block Resection

EVERETT J. GORDON, M.D.*

Malignant bone tumors are encountered infrequently in private practice. The prognosis usually is grave, despite such heroic surgical measures as amputation, disarticulation or hindquarter amputation, which generally are recommended in malignant bone tumors. This report concerns a rare type of bone tumor of relatively low malignancy in which conservative surgery has yielded an excellent result. It demonstrates that a well-planned block resection of a limited tumor can produce a much more satisfactory result than an amputation, with its profound social and economic implications.

Parosteal osteogenic sarcoma was first described as a distinct clinical entity by Geschickter and Copeland[3] in 1951, when they acquainted the Southern Surgical Association with 16 cases. They described the neoplasm as parosteal osteoma with a histologic picture resembling myositis ossificans, but with more intimate relationship to bone and of a graver prognosis. They originally described the tumor as occurring in both benign and malignant forms, the latter predominating, and occurring usually in an adult of 20 to 40 years of age. The tumor was believed to originate in extra-osseous connective tissues, possibly derived from remnants of the primitive limb bud. Previously others had described this type of tumor as an atypical myositis ossificans, but Geschickter and Copeland clearly demonstrated distinct differences between the two entities in roentgenographic, gross and microscopic appearance. The juxtacortical position, propensity for occurrence in the lower third of the femur and the upper portion of the humerus, and the tendency to local recurrence with possible distant metastases after simple resection were all stressed as characteristic of this particular tumor. Of 13 cases, 7 recurred after local excision, of which 4 died of pulmonary metastases. It was their recommendation that cases that were positively identified histologically should be treated by amputation.

In 1953, Dwinnell[1,2] reviewed 2,000 primary bone tumors at the Mayo Clinic, including 400 cases of osteogenic sarcoma, and found 15 instances of juxtacortical osteogenic sarcoma: 10 were in the distal femur, 3 in the humerus and 2 in the tibia, with an age distribution of 10 to 39, mostly in the fourth decade, and no sex differences. This indicates an incidence of 0.8 per cent of all bone tumors and less than 4 per cent of all osteogenic sarcoma. The most frequent presenting symptom was a mass, present for one or more years, usually without pain, but occasionally with mild pain of short duration, with occasional loss of motion in an adjacent joint due to the mechanical in-

* Washington, D.C.

terference of the protruding mass. In no instances were there evidences of fever, anemia, weight loss or leukocytosis.

The various names given this particular tumor—periosteal osteogenic sarcoma, parosteal osteoma, juxtacortical osteogenic sarcoma and parosteal osteogenic sarcoma—apply to an ossifying type of lesion with a broad base occurring on the surface of long bones without penetration into the medullary cavity. The roentgenologist may confuse this tumor with an atypical osteochondroma, despite the fairly typical appearance of an ossifying lobulated mass, juxtacortical, attached to the metaphyseal area or the adjoining epiphyseal region, containing scattered areas of lesser density throughout the bony mass, representing cartilage, trapped fascia and muscle. The tumor mass usually grows along the shaft or circumferentially, without invading the medullary cavity primarily. The underlying cortex is irregular and thickened but shows little reaction of destruction. Later, after one or more unsuccessful attempts at local excision, the tumor may undergo frank sarcomatous changes and show sclerosis, osteolysis and destruction of the cortex and the medullary cavity. Pathologic fractures are uncommon, as the tumor is densely ossified.

Grossly, the growth has an irregular lobulated surface with absence of a cartilaginous cap as found with an osteochondroma; it consists of bony tissue, plus softer areas of infiltration by cartilaginous and fibrous tissue, all encapsulated in an adherent fibrous tissue sheath It is a tumor arising from periosteum and retaining its bone-forming tendencies: the cut section shows the mixed appearance caused by bone, fibrous tissue and cartilage, with bone predominating. In a primary tumor, no necrotic areas are found, but with recurrent tumors of higher malignancy such areas may be noted.

Microscopically, a typical section will show nonmalignant bone spicules with interstices filled with atypical spindle or polyhedral cells, occasional or numerous mitotic figures, no fat cells, but variable amounts of new osteoid distributed among the proliferative spindle cells and occasional calcification in the larger zones. Also noted are cartilaginous islands reflecting various degrees of malignant changes in their peripheral cells, which blend with the interlacing screens of spindle cells. Occasional benign giant cells and areas of chondrosarcoma are also noted frequently. In recurrent tumors, advanced malignant features are found more frequently. Almost all cases are of low malignancy, usually of Grade 1 of Broder's classification.

The diagnosis of a juxtacortical lesion is not always clear, and other lesions besides parosteal osteogenic sarcoma should be considered in diagnosis. Osteogenic sarcomas may present some resemblance initially, but usually they have medullary involvement, accompanied by pain, osteolysis of the cortex, and a periosteal sun-ray reaction by x-ray. Exostoses and osteochondromas are confused frequently with juxtacortical osteogenic sarcoma, as occurred in the case now being presented, but these lesions have orderly trabecular patterns within the tumor and no scattered radiolucent areas, as no fascia or muscle is included in their masses. Small tumors in tendon sheaths or ligaments, known as sesamoidomas, can usually be differentiated by their location and size. Parosteal osteogenic sarcoma has long been confused with heteropic ossification or myositis ossificans However, the latter lesions always have a history of trauma and demonstrate laminated ossification parallel to the shaft with a definite noninvolved area separating it from the underlying bone, demonstrating its lack of continuity with the bony cortex They also lack the new osteoid found in juxtacortical osteogenic sarcoma. The recently organized callus found after fractures can be confused with these tumors, but the history is diagnostic. Periosteal chondromas and chondrosarcoma lack the prominent bony and fibrous components of juxtacortical osteogenic sarcoma.

An important diagnostic point in recognition of recurrent tumors after local excision is the tendency of this particular type of *bone tumor to recur in the image of its* primary tumor, often closely simulating its original appearance.

## TREATMENT

Parosteal osteogenic sarcoma is recognized as the most curable of all malignant bone tumors, although frequently the diagnosis is missed. The form of treatment recommended most generally is amputation. Stevens,[5] who continued the review of Mayo Clinic cases, finding a total of 19 cases between 1905 and 1955, recommended amputation but noted that local extirpation could occasionally be performed. He warned against seeding of the tumor by biopsy, and advised roentgenographic examination before closing the wound after local resection. To ensure complete extirpation of the tumor, especially with circumferential involvement of the shaft, where a small remnant of tissue could easily be overlooked. All authors strongly emphasize the dangers of inadequate removal of this tumor, noting the strong tendency to local recurrence in a more malignant form with subsequent pulmonary metastases. Almost all the reported deaths occurred from pulmonary metastases in recurrent tumors. Dwinnell,[1,2] Geschickter and Copeland,[3] and Sammons[4] all recommend amputation once a histologic diagnosis of juxtacortical osteogenic sarcoma is made. In Dwinnell's series, 9 of 10 cases treated by local excision had recurrences, and, of these 9, 4 subsequently died of metastases.

Although amputation is undoubtedly the safest treatment, it would seem that certain carefully selected cases can be treated initially by less radical surgery, inasmuch as this tumor has a well-established low degree of malignancy. In localized tumors, in which sufficient uninvolved bone exists to permit adequate block resection with a periphery of normal bone in the resected tumor block, a



Fig. 1. August, 1955, preoperative roentgenogram. Cortical mass, metaphysis, lower femur, no medullary involvement.

good result can often thus be achieved. However, it is stressed that in the majority of cases, amputation is mandatory and lifesaving, and great care should be exercised in selecting cases for more conservative surgery. The advantages accruing to the fortunate patient who can be treated by block resection are quite obvious and may materially affect his whole social and economic life.

## CASE REPORT

A 16-year-old white female was first seen on August 22, 1955, referred for consideration of amputation for a bone tumor. She had noted *swelling on the anterior surface of her left lower* thigh for the past year but with severe pain of [illegible]

April, 1955.

*Examination revealed a firm swelling of the* left lower thigh immediately above the patella, with considerable soft-tissue swelling and tenderness about a firm bony mass. The left knee measured 1 inch greater in circumference than the right at the suprapatellar level. The patient *had full knee flexion to 25° but lacked 5° of full* extension of the left knee. There was mild atrophy of the left thigh, measuring ½ inch less in circumference than the right at the mid-point. The knee ligaments were intact, and there was normal motion in the left hip and ankle. Labora-

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FIG 1. August, 1955, preoperative roentgenogram Cortical mass, metaphysis, lower femur, no medullary involvement.

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FIG. 2. Gross specimen, 6 inches long, showing irregular lobulated surface with scattered areas of decreased density. Normal cancellous bone immediately above rule.

FIG. 3. (*Top*) Photomicrograph (×30, hematoxylin and eosin stain) showing parosteal osteogenic sarcoma; pseudopodial pattern of maturing chondrocytes and young osteocytes. (*Bottom*) Photomicrograph (×30, hematoxylin and eosin stain) showing tumor breaking through cortex with resultant periostitis.



FIG. 4. December 14, 1955, postoperative roentgenogram. Note bony defect, partially filled with two cortical bone grafts extending into femoral condyles. Knee joint intact.



FIG. 5. August 24, 1956, 1 year after operation. Excellent consolidation of bone grafts. No evidence of recurrence.

tory data revealed no significant abnormal changes in the urine or the blood count. Serologic test for syphilis was negative; the girl had an 89.3 per cent hemoglobin.

Roentgenologic studies of the left knee (Fig. 1) reported a bony mass, the size of a small lemon, on the anterior aspect of the left femur, slightly irregular and jagged, with raised periosteum, not involving the knee joint, diagnosed as a probable benign tumor, possibly an aberrant type of osteochondroma.

On August 26, 1955, at the George Washington University Hospital, a block resection of the tumor mass of the left lower femur, plus a peripheral zone of normal bone, was performed under pneumatic tourniquet. The mass extended down to the supracondylar spongy bone of the femur. Complete removal involved resection of the anterior two thirds of the cortex, leaving only the posterior third of the femur intact. A thick fibrous cap of the tumor was also removed (Fig. 2). Two large cortical bone grafts were inserted in the defect created and supplemented by *bone burger*, ground cancellous rib bone, generously donated by the tissue bank of the National Naval Medical Center, Bethesda, Md A long-leg cast immobilized the leg from the groin to the toes. The postoperative course was uneventful, and the patient was discharged from the hospital 1 week after surgery —September 2, 1955.

The local pathologist diagnosed the tumor as *ossifying ecchondrosis*, but a gross section, forwarded to the Armed Forces Institute of Pathology, was reported by Dr. Lent C. Johnson as follows:

"Very typical, well-developed parosteal sarcoma with adequate removal, with good margin and normal bone in all directions, together with almost entire cavity underlying it.

"The diagnosis of sarcoma is based upon two aspects: first, that the pattern of varied types of skeletal tissue within the mass is in a rather chaotic arrangement; and second, that the most actively growing cells are on the surface with small nodules budding off therefrom like pseudopods extending out from the main mass of the tumor. Cytologically, these surface cells have the characteristics of any malignant tumor despite the fact that the rest of the tumor quiets down and differentiates enough to make certainty of malignancy difficult. The whole bone sections make it very clear that the tumor has



FIG. 6. June 5, 1959. Solid bony union with mild residual bony defect anteriorly. No recurrence. Remolding of bone architecture noted.

arisen on the surface of the bone and is just beginning to break through the cortex after having reached a considerable size on the surface." (Figs. 3 & 4.)

Two months after surgery—on October 24, 1955—the cast was removed, the wound was well healed, and roentgenograms showed the grafts integrating with early callus formation. Cautious physical therapy was begun to restore knee motion, and the patient was allowed to be on crutches without weight-bearing. On November 9, 1955, partial weight-bearing with crutches and an ischial caliper brace was begun; more intensive physical therapy was instituted with whirlpool baths, Delorme passive resistive exercises, and bicycle exercises to regain knee motion and quadriceps strength. There was considerable weakness in the quadriceps, but this gradually improved, together with the range of knee motion Crutches were discarded on December 14, 1955, 4 months after operation, at which time roentgenograms (Fig. 5) showed the bone grafts well integrated and intact with definite callus formation present at the ends of the grafts At this time, the patient had a range of motion in the left knee of 62° to 170° with difficulty in full extension, due to atrophy and adherence of the vastus intermedius muscle to the underlying bone at the resected area.

On February 29, 1956, 6 months postoperatively, motion of the left knee was from 50° to 170° compared with 35° to 175° on the right At this time firm bony union of the cortical strut grafts was noted by roentgenogram, and the ischial caliper brace was discarded One year postoperatively—August 27, 1956—the range of knee motion was from 50° to 177°, adequate for excellent function, and the girl was walking without a limp or instability and had no quadriceps weakness. There was no lymph-node enlargement, and roentgenograms showed the grafts well consolidated with firm union (Fig. 6)

On May 26, 1957, 21 months after surgery, the patient had a range of motion of 35° to 180°, equal to that in the right knee, with good stability, no crepitus, no atrophy and a normal gait. On June 5, 1959, 4 years postoperatively, she was walking without a limp, with no complaints, good quadriceps strength, no local masses, no tenderness, no swelling. There was no evidence of any metastases or local recurrences, either clinically or by roentgenogram (Fig. 7).

The patient was married, had a normal pregnancy and delivery, and was living an unhandicapped, happy life.

## COMMENT

It is believed that the term *parosteal osteogenic sarcoma* is a more descriptive one for a definitely malignant lesion than the original term of *parosteal osteoma,* applied by Geschickter and Copeland. The presence of a firm bony mass, near a joint, of many years' duration with little or no symptomatology does not necessarily imply a benign osteochondroma or exostosis, as commonly misdiagnosed. Although relatively rare, such a bony mass may represent the malignant type of lesion under discussion. If diagnosed sufficiently early, before extensive malignant changes have occurred, successful well-planned block resection can be performed. It is not uncommon for such a lesion to go undiagnosed for one or more years, but block resection may still be possible if the tumor is of limited size without circumferential growth. If the tumor has a circumferential position, amputation usually is the only choice. Certainly, in all cases of local recurrence, immediate amputation should be performed.

## SUMMARY AND CONCLUSIONS

1. Parosteal osteogenic sarcoma is a relatively rare bone neoplasm of low-grade ma-

lignancy, with characteristic roentgenographic and clinical findings sufficient for differential diagnosis from other types of bone tumors. However, a definitive diagnosis must rest upon histologic examination.

2. It is this author's opinion that small parosteal lesions should be treated by block resection instead of biopsy, as biopsy may lead to seeding of the tumor, and, besides, a definite diagnosis cannot be made by frozen sections at the time of surgery. Later, if parosteal osteogenic sarcoma is diagnosed histologically, no further surgery will be required in many cases, depending on the extent of malignant cytologic changes and thoroughness of tumor removal. Roentgenographic check on the operating table for complete extirpation of neoplastic tissue may be a valuable therapeutic aid.

3. Amputation is the treatment of choice in most instances of parosteal osteogenic sarcoma. However, block resection with a generous margin of normal bone should offer an excellent result in carefully selected cases with small tumor growths of low malignancy without circumferential involvement of the cortex.

4. Each bone neoplasm must be treated as a separate clinical-pathologic entity and an individual decision made regarding treatment.

5. A case is presented in which block resection of a parosteal osteogenic sarcoma and grafting of the bony defect created produced an excellent result without evidence of local or systemic metastases 4 years after surgery.

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## Parostee Sarcoma Osteogene; Tractamento per Resection de Bloco

*Summario in Interlingua*

defecto ossee per graffos homogenee. Le diagnose pathologic original habeva essite "ecchondrosis ossificante." Subsequentemente le correcte diagnose de parostee sarcoma osteogene esseva facite per le Instituto de Pathologia del statounitese Fortias Armate. Isto illustra le facto que un section congelate es sin valor in tal casos, proque mesmo un plenmente processate specimen o un biopsia pote esser interpretate incorrectemente per un pathologo qui ha solmente sporadic contactos con tumores de osso, con le resultato possibile de un tractamento inadequate. Le patiente esseva capace de ambular sin crucias quatro menses post le operation. Tres annos e medie plus tarde, illa habeva le complete function del extremitate in question e nulle signos de metastases.

Un resection complete de bloco in loco de biopsias es recommendate in cautemente seligite casos de micre crescentias tumoral que non monstra un affection circumferential del cortice. Amputation es indicate in omne casos de recurrentia e es le tractamento de election pro extense crescentias de parostee sarcoma osteogene.

## SECTION III

# ITEMS

# 19

# The Operative Treatment of Diaphyseal Fractures in Children

GUILLERMO DE VELASCO Y POLO, M.D.*

Many different factors have made us depart from the usual conservative methods of treatment of fractures in children. These factors are present-day advances in the methods of treatment of fractures, the control of infections by antibiotics, the lack of hospital beds, and socioeconomic problems peculiar to Mexico. They have influenced our criteria, so that now we prefer a surgical technic in a certain type of fracture that produces results equal to, or better than, those obtained by conservative management.

The statistics presented in table on this page cover operations performed from 1943 to date in the Orthopaedic Services I and II, and, since 1945, in the Shriners' Service and the Emergency Service of the Children's Hospital.

In the Emergency Services at the Children's Hospital there is no special traumatology service. Because of the great number of patients with locomotor disturbances, the 120 beds assigned to the orthopaedic services are reserved for these patients, and this makes it almost impossible to obtain a bed for a patient with skeletal trauma. Only on very few occasions do the surgery services grant us beds for such patients. Therefore, we find ourselves confronted by the serious hospitalization problem of children with diaphyseal fractures, especially of the lower limbs, who, if we wished to treat them by conservative methods, would occupy our beds for no less than 30 to 40 days. For this reason one encounters a certain reluctance on the part of the chiefs of services to admit them. If they were to be admitted, it would mean that in a few months we would tie up a large number of the already small number of beds available for solving their own problems; i e., the hundreds of

OPERATIONS PERFORMED FOR DIAPHYSEAL FRACTURES IN CHILDREN FROM 1943 TO 1958 (Age, 2 to 14 Years; Average Age, 6 Years, 6 Months)

| | No. of Cases | Percentage |
|---|---|---|
| Femur | 260 | |
| Exposed femur | 1 | |
| Forearm | 134 | |
| Elbow | 59 | |
| Tibias | 5 | |
| Humerus | 14 | |
| Clavicle | 17 | |
| Hips (diaphysectomies) | 64 | |
| Metatarsals | 6 | |
| Metacarpals | 3 | |
| Phalanges | 2 | |
| Total | 565 | |
| Mortality | | — |
| Infections | | — |
| Delayed consolidations | | 0.33 |
| Pseudarthrosis | | — |
| Pathologic cases | | 0.63 |

* Mexico City.

patients on their waiting lists. On the other hand, if we consider the expense of treatment, we can see, by means of simple multiplication, that 1 patient hospitalized for 30 days would cost the hospital about 2,000 pesos. For the same patient treated by surgical means it would be no more than 100 or 200 pesos. Thus, the cost per patient is reduced to less than a tenth, and the number of patients cared for is multiplied by 10.

We have outlined briefly the hospital problem. Let us now consider the social problem. As is well known, the Children's Hospital is at present about the only one of its kind in the Republic of Mexico, so that, in spite of its being unable to meet the requirements of the Federal District alone, it has to take on all the patients who are sent to it from the various parts of the Republic. Boiled down, it means that we have only 120 beds for the surgical treatment of orthopaedic patients in a nation of over 20 million inhabitants.

The majority, if not all, of those who enter our services are in very low socioeconomic circumstances. Therefore, we have no confidence in sending them away for treatment at home. It would mean giving them various traction apparatuses and plaster casts that would not be used correctly, and the majority of patients would end up with badly consolidated fractures, limb shortening, pseudarthrosis, etc. Ultimately this would constitute a very serious problem, both for the hospital and for the patient and his family These are the factors that led us to change our thinking regarding the treatment of diaphyseal fractures. We now look to surgery for their treatment.

Which children do we treat by this method? When the fracture is capable of being reduced by closed methods—i.e., the frequent greenstick type of fracture—and we can undertake it with confidence, we do so and discharge the patient rapidly. On the other hand, when we know that the reduction will be hard to maintain—i.e., the transverse fracture of the femur or the double fracture at the middle third of the forearm—we decide on a surgical solution. There are some cases, like the spiral or the oblique fractures of the tibia, with great displacement of the fragments, which, even though they could be treated conservatively by various methods in the hospital, we deal with surgically. This is done in order to discharge the patient quickly and avoid the probable poor care by other methods at home. Compound fractures are treated according to classic methods, conservatively if the wound is infected or if over 8 hours have elapsed since trauma. Antibiotics and antitetanus toxin are administered, and, when all danger of infection has passed, convenient reduction and fixation are carried out. In some cases, with wounds seen within 6 hours after they occurred and having a good clinical appearance, we hazard operation. Thus the problem is resolved in a single intervention and is controlled by antibiotics. It is to be noted that special circumstances have led us to intervene in compound comminuted fractures. In these cases we have solved the problem surgically with excellent results.

Which surgical technics do we use? In our early days we used wire transfixion and, in some cases, bone plates. We have since discarded the latter, and now only the former is used. The wire we use to reinforce a fixation or to fix small fragments over which one has little control. Reviewing our statistics, we see that the most frequent fractures of the operative type are of the shaft of the femur. In diaphyseal transverse fractures we carry out Küntscher-type intramedullary nailing. We never carry out the closed method technic conceived by the author because we believe, with Colonna, that it is contraindicated Because of the small caliber of the intramedullary canal and of the difficulty of the method, we do not feel that it is safe to expose a child to a long anesthesia and large-dosage roentgen-ray radiation. Instead, we open the fracture site and pass the pin up through the trochanter until the distal end of the pin is flush with the fracture. We

reduce the fracture and bring the pin down the distal fragment, stopping short of the growth cartilage. We do not use laminated or diamond-shaped pins, considering that, aside from their high cost, they can produce serious lesions in the medullary canal. Steinmann pins ensure excellent fixation: the quality of the steel alloy is insuperable; and they come in different sizes to meet the individual case. A spica cast is used in the majority of cases, although it is feasible to leave the child with a small plaster-of-Paris boot to which a small wooden crosspiece is affixed. In this manner we avoid rotation at the fracture site.

When the roentgenograms indicate satisfactory union, we withdraw the pin through a small incision in the gluteal region. With this method it is possible to get the patient on his feet at an early stage and mobilize the joints temporarily to avoid rigidity.

In oblique tibial fractures, with displacement of the fragments, we do an open reduction and fix it with a screw and a plaster cast for a period of 6 to 8 weeks. We do not favor intramedullary nailing of the tibia in children.

In transverse fractures of the forearm, we first pin the ulna by a back-and-forth technic, then pin the radius, passing the pin through the distal radial epiphysis, slightly posterior to the abductor tendon. It is feasible in these cases, when one of the bones is comminuted, to carry out an intramedullary pinning on one of the bones and fix the other fracture with wire transfixion.

In the humerus we first pass the intramedullary pin through the distal fragment, so that the pin comes out through the trocheal humeri. Reduction is carried out, and the pin then is passed up the proximal fragment. The pinning is not done through the head of the humerus, because it is a more difficult technic and it is possible to injure the joint or the growth cartilage.

As one can see from the statistics, in children it is rarely necessary to operate for fractures of the clavicle. We operate on the clavicle only when the separation between the fragments is great. In the transverse fracture it is easy to do the pinning by using a larger size Kirschner wire. This is passed first through the distal fragment, coming out at the posterior aspect of the shoulder, and is then passed into the proximal fragment. In oblique fractures we use an X-type of transfixion with stainless-steel wire.

In the phalanges we use intramedullary pinning with Kirschner wires, passing the wire first through the distal fragment and then up through the proximal fragment.

On reviewing our statistics, we find that, over a period of 12 years, the operative method of treating certain types of fractures in children has given us good results in all cases. The method has been adopted because of this and of the socioeconomic and hospital factors enumerated.

## Le Tractamento Chirurgic de Fracturas de Diaphyse in Juveniles

*Summario in Interlingua*

Es discutite varie factores que argue in favor del tractamento chirurgic de fracturas de diaphyse in juveniles. Tal factores es le problema de spatio in le hospitales, problemas economic e de familia, e etiam le cambiamentos que ha occurrite in le chirurgia depost le advento del *antibioticos*. Super le base del experientia con circa cinque centos casos, tractate inter 1943 e le data presente al Hospital Infantil de Mexico, tanto in le servicio de urgentia como etiam in le servicio del Shriners, le conclusion es presentate que le uso de simple technicas, con solmente clavos de Steinmann o vites, permitte le obtention de cento pro cento de bon *consolidationes a longe vista durante que le* supra-mentionate problemas es resolvite o evitate.

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# 20

# Doctors Encouraged To Utilize the Latest Anatomic Terminology (Nomina Anatomica)

W. COMPERE BASOM, M.D.*

At the meeting of the Board of Governors of the American Fracture Association held during the convention of the American Academy of Orthopaedic Surgeons in Chicago, Ill., on January 25, 1959, it was voted unanimously to encourage the distribution and the use of the latest anatomic terminology. The terms adopted by the International Nomenclature Committee, approved by the Fifth International Congress of Anatomists at Oxford, 1950, and eventually ratified at the world meeting in 1955, were considered to be an excellent basis of communication among doctors throughout the world. The Board also felt that their use would tend to better Latin-American relations. Dr. Juan Farill, the regional vice-president from Mexico, supported the new terminology enthusiastically and urged th members to brush up on the new terms.

The firm of Williams and Wilkins, wh have distributed free the reports of th Nomenclature Committees and a complet list of the new terms, will be urged to publisl this booklet at a fee, and the Board vote that the Association purchase 500 copie for distribution to their own members.

It is hoped that other medical organiza tions also will take steps to encourage th use of the latest terminology. Actually, thi terminology is based on the *Basle Nomin Anatomica*, which is on a Latin basis. Th terms are not difficult; they are merely ol friends in slightly different dress.

Now, when the accent is on better international relations, this would seem to be a big step forward.

* El Paso, Tex

# 21

# Coccidioidomycosis of the Hip Joint

## Case Report

LOUIS W. BRECK, M.D., LESLIE M. SMITH, M.D., AND ROBERT E. HAAS, M.D.*

In the arid semitropical region of the border between the United States and Mexico, coccidioidomycosis is probably seen more frequently than anywhere else, except in the San Joaquin Valley of California. However, it is still relatively rare. Although we are constantly on the lookout for this disease, the case to be discussed was not diagnosed until late in its course. The dermatologist (L.M.S.) was not called in until the diagnosis of coccidioidomycosis had been made. The evolution of the case will be seen in the report that follows.

Over a period of years, the two senior authors (L.W.B. & L.M.S.) have seen a number of cases of osseous coccidioidomycosis, many of them without demonstrable systemic involvement. The majority have involved the lower extremity. They have responded only fairly well to any treatment, and many have remained as chronic draining sinuses or have required amputation. However, the mortality from the disease has been surprisingly low in those cases without severe systemic involvement. In 1951, we collected a series of cases that clinically were Madura foot, several of which proved to be coccidioidomycosis. The majority were actinomycotic in origin. The present case is interesting and unusual from several aspects, which will be commented upon later.

* El Paso, Tex.

## CASE REPORT

A 76-year-old Mexican male fell on October 8, 1953, and fractured the neck of the left femur. Within a few days an open reduction with fixation by means of screw and nail was accomplished in Juarez, Chihuahua, Mexico. Afterward, however, drainage developed at the operative site. This continued, and on July 8, 1954, the man again underwent operation, at which time a Fred Thompson Vitallium-type prosthesis* was inserted. After this procedure, which also was done in Juarez, the patient returned to his home at Villa Ahumada, a rural community in the state of Chihuahua. He stated that subsequently the drainage became more profuse.

He presented himself to us as a referral on December 15, 1954, because of drainage and pain in his hip with loss of motion and consequent inability to walk. Examination at that time revealed evidence of inflammation over the lateral aspect of the left hip with a sinus which, according to the patient, drained from 4 to 6 cc. of pus each day. There was practically no hip motion. Roentgenographic evaluation demonstrated the acetabulum to be practically dissolved with the prosthesis well seated in the femoral shaft, but the head of the prosthesis appeared to be partially dislocated (Fig. 1).

It was felt that conservative treatment with antibiotics was the method of choice. Accordingly, this was instituted through his referring physician but with little effect. Drainage con-

* Thompson, F. R.: Two and one half years' experience with a Vitallium intramedullary hip prosthesis, J. Bone & Joint Surg. 36-A:489-500, 1954



FIG. 1. Roentgenographic appearance with active coccidioidomycosis present and before removal of Fred Thompson Vitallium hip prosthesis.



FIG. 2. Roentgenogram made 6 months after removal of the hip prosthesis. At this time drainage had almost stopped. It did soon afterward.

tinued until, finally, on August 6, 1955, the patient again was operated upon, and the prosthesis was removed. At operation considerable difficulty was encountered in removing the prosthesis because of scarring. The sinuses, the [illegible] tabulum. Cultures and sections of the tissue subsequent to surgery revealed a coccidioidal infection of the hip joint and adjoining structures The patient was referred to the dermatologist, who treated him conservatively with penicillin, sulfonamides and testosterone. By May, 1956, 6 months after operation, all sinuses had closed and he appeared to have recovered. Roentgenograms showed considerable new bone formation about the joint and no evidence of destruction (Fig. 2). He was allowed up on crutches in February, 1956, and by June he was getting about satisfactorily with no evidence of a recurrence.

The patient was last heard from by letter in February, 1958, when he was alive and well with no recurrence of drainage or evidence of recurrence of the disease.

## COMMENT

This case presented to us as postoperative infection of the hip joint. Simple culture of the wound showed the presence of *Staphylococcus aureus*, which is what one ordinarily would expect in such a case. Originally, no special examination for coccidioides was done, as it was not even suspected to exist

Removal of the large Vitallium hip prosthesis was advised, as this is the best way to stop draining infections in the hip joint in cases of this type after everything else has failed, and this was done.

The authors suspect that the coccidioides got into the patient's wound while he was at home a considerable time after the original drainage started. It is possible that while he was being dressed for a simple pyogenic infection of the hip region, the coccidioides entered the sinus either from the fingers or the dressings used on the wound. The man lived on a farm in a cotton field 75 miles south of El Paso, Texas, in a relatively primitive section of northern Mexico. The draining wound was dressed at regular intervals by the patient himself, some member of the family or some unskilled household attendant. The last-mentioned did most of the

dressings, and sterile technic was not employed. The dressings were done in a rather crude manner. The authors do not believe it likely that the original operative infection could have contained the organism. It is hard to see how this organism could withstand the sterilization employed with surgical technic or how it could be present in the operating room. However, this is purely conjectural.

At the time of operation when the hip prosthesis was removed, the acetabulum looked most unusual, in that about half of the acetabular area contained a thin granulomatous type of growth that was dark green in color. At operation, sinuses were found to be running in several directions, and there were several extensive pockets.

After the hip prosthesis was removed and the diagnosis was made in the hospital a few days after the operation, the dermatologist (L.M.S.) was called in to treat the case, and treatment was administered vigorously from then on. After a considerable period of time, a very satisfactory cure was obtained, as outlined in the case report. In view of the very extensive infection and the patient's age, it was considered to be somewhat remarkable that a satisfactory result was obtained.

At the present time the patient is well. His hip stopped draining long since. It is somewhat painful because of its anatomic condition, with no head and neck and only scar tissue in the area. The patient walks well with crutches and can walk with a cane. He now wants to have something more done for this hip to make him less disabled, but the authors do not contemplate doing anything further, for they consider themselves lucky to have a live patient with no drainage and with an apparent complete cure of his coccidioidomycosis.

## MEDICAL TREATMENT (L.M.S.)

While there is no recognized specific treatment for coccidioidomycosis at this time, persistent general and local treatment will often be rewarding if the disease is not severe and generalized, and if the patient's resistance to the organism is good, as indicated by a positive coccidioidin test. The patient under discussion had a negative skin test and no evidence of systemic disease. The large draining sinus was injected with 10 per cent copper sulfate solution every 2 or 3 days for the double purpose of killing as much of the coccidioidal and staphylococcal infection as possible and of promoting fibrosis in the sinus. This caused some pain and temporarily increased the drainage, but the latter diminished gradually. Penicillin injections, Gantrisin orally and methyltestosterone 150 mg. daily were given, the penicillin and the sulfonamide to aid principally in the control of the secondary infection. Lamb* has treated some cases of coccidioidomycosis with sulfonamides and testosterone with apparent benefit. Testosterone seems to have some fungicidal action. After 3 months the material draining from the sinus, though less in quantity, still contained a few coccidioidal bodies. Lugol's solution, 15 to 75 drops daily, was added to the treatment, and the copper injections were discontinued. By May 1, 1956, 9 months after starting treatment, the sinus had closed, there were no apparent signs of inflammation, and the patient regarded himself as cured. Reports from him by mail at intervals have all been good, and for a year and a half he has had no signs of the disease. Unfortunately, the patient has not been in of late for an examination, but a recent letter states that there has been no further evidence of the disease.

Coccidioidomycosis is a capricious disease, subject to remissions and exacerbations. Therefore, it is difficult to evaluate treatment, and one hesitates to report a cure of even a localized lesion, but the disease in this case remaining for 2½ years without signs of symptoms, we feel that the condition

* Lamb, J. H.: Combined therapy in histoplasmosis and coccidioidomycosis; methyltestosterone and meth-dia-mer-sulfonamides, A.M.A. Arch. Dermat. & Syph. 70:695-712, 1954.

has been walled off and arrested by fibrosis, if not cured. A negative complement fixation test and a final negative roentgenographic examination would be further criteria of cure, and these we still hope to obtain, but it is difficult to get the patient to make the trip to El Paso when he sees no signs of disease.

## SUMMARY

A case of coccidioidomycosis of the hip joint and hip region is presented. This infection ensued following a series of surgical events: a fracture of the hip, a hip nailing with an operative infection and the insertion of a Fred Thompson Vitallium hip prosthesis for a nonunion of the fracture of the hip. The patient then was referred to us, and the prosthesis was removed. Following this last procedure a coccidioidomycosis was found to exist.

The infection with coccidioides is believed to have been due to the introduction of organisms into a draining sinus when the wound was dressed at home on a cotton farm 75 miles south of El Paso, Texas, in Old Mexico.

Besides removing the prosthesis, all the granulation tissue and the infected tissue that could be found was excised. This included several pockets and sinuses.

The infection with coccidioides was not discovered until a few days after operation, when the bacteriologic laboratory reported it. It had not previously been suspected, instead, an ordinary *Staphylococcus aureus* infection was thought to be present, since this organism had been found in preoperative cultures.

Extensive treatment by the dermatologist (L.M.S.) with large doses of penicillin, sulfonamides and testosterone after about 9 months resulted in a complete clinical cure.

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